

Royal College of Paediatrics and Child Health

MRCPCH PART II EXAMINATION

PAPER 2

1. Complete the following:

Your full name (BLOCK LETTERS)

.....

RCPCH Number

Signature

2. Please check that your surname (family name) is correct on the **Answer Sheet**.

Do not write anything on the Answer sheet other than your response to each question.

3. Answer all of the questions.

4. It is strictly forbidden to talk to, read the work of, or attempt in any way to communicate with, other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work. The College has tools which can identify copying of answers or collusion between candidates to share answers. In any situation the suspicion of guilt falls upon both parties until it can be proved otherwise. Breaches of these instructions, or misbehaviour in any other way, including continuing to write after the allotted time, may lead to suspension from the examination at the discretion of the invigilators. Serious breaches, such as cheating or colluding to gain advantage, could incur permanent suspension from College examinations.

5. Copyright law protects examination questions and the intellectual property of their authors. The unauthorised use of questions is a breach of copyright law.

6. Time allowed: 1 hour.

Questions 1 and 2

An 8-month-old male infant is referred because of non-bilious vomiting. His general practitioner (GP) had seen him frequently for constipation over the last few months.

Examination reveals a thin, non-dysmorphic infant weighing 6.8kg (1st centile). He has a scaphoid abdomen and his capillary refill time is three seconds. General examination was otherwise unremarkable.

Routine blood results:

Hb	12.2 g/dl
WBC	$13 \times 10^9/l$
neutrophils	$9.4 \times 10^9/l$
lymphocytes	$3.6 \times 10^9/l$
Platelets	$373 \times 10^9/l$
plasma sodium	154 mmol/l
plasma potassium	3.8 mmol/l
plasma urea	6.0 mmol/l
Urine microscopy	no red cells no white cells no casts
Urine osmolality	180 mOsm/kg

This question continues on the next page.

1. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

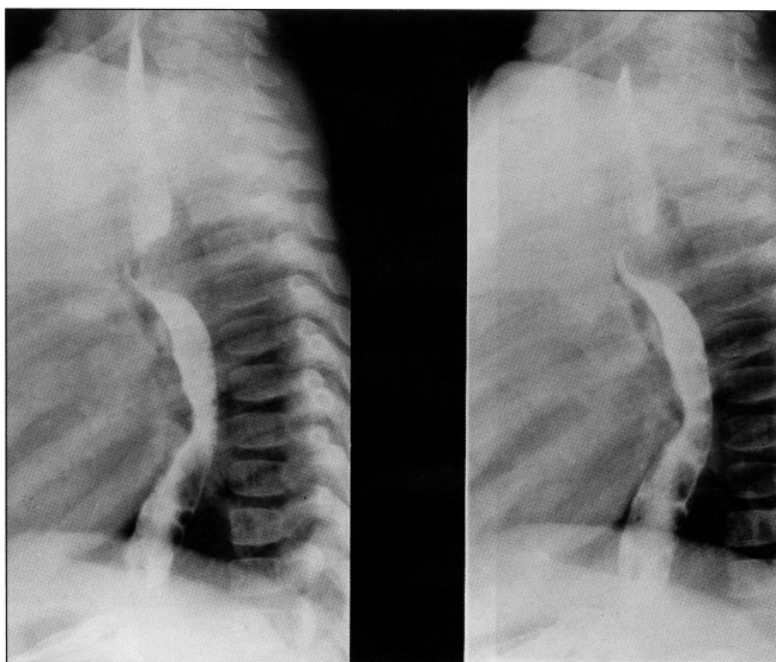
- A chronic renal failure
- B diabetes insipidus
- C diabetes mellitus
- D gastro-oesophageal reflux
- E Hirschsprungs' disease
- F renal tubular acidosis
- G urinary tract infection

2. Which of the following is the most appropriate test to confirm the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A blood sugar
- B oesophageal pH study
- C plasma aldosterone level
- D plasma creatinine
- E plasma osmolality
- F rectal biopsy
- G response to DDAVP
- H urinary pH
- I urinary steroid profile

Questions 3 and 4



Q3

A 7-month infant presented with two episodes of stridor

3. What two features does the x-ray show? (4 marks)

SELECT TWO ANSWERS ONLY

- A enlarged heart
- B gastro-oesophageal reflux
- C impaired oesophageal motility
- D mid-tracheal compression
- E posterior oesophageal compression
- F oesophageal stricture
- G upper lobe consolidation

This question continues on the next page.

4. What is the most likely diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A achalasia
- B bronchomalacia
- C gastro-oesophageal reflux
- D paravertebral abscess
- E pulmonary stenosis
- F tracheal foreign body
- G tracheo-bronchial aspiration
- H vascular ring

Question 5



Q5

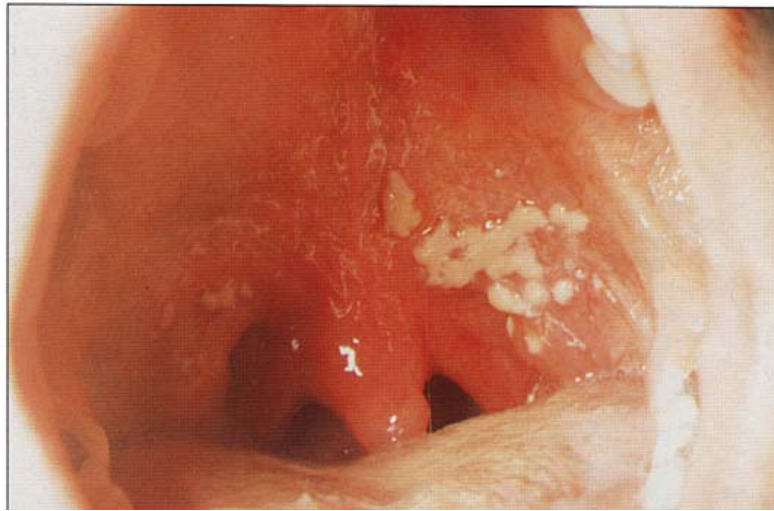
This x-ray was taken of an 18-month-old child on admission with impaired movement of the left arm.

5. What are the two radiological diagnoses? (2 marks)

SELECT TWO ANSWERS ONLY

- A fractures of radius and ulna
- B lead poisoning
- C mucopolysaccharidosis
- D non-accidental injury
- E osteogenesis imperfecta
- F osteomyelitis
- G osteopetrosis
- H pseudohypoparathyroidism
- I rickets

Questions 6 and 7



Q6 & Q7

This teenager was 16 years old

6. What is the lesion on the soft palate? (1 mark)

SELECT ONE ANSWER ONLY

- A drug reaction
- B Epstein-Barr virus infection
- C herpes simplex infection
- D oral candidiasis
- E staphylococcal infection
- F streptococcal tonsillitis

This question continues on the next page.

7. Give three possible predisposing factors. (3 marks)

SELECT THREE ANSWERS ONLY

- A anti-fungal therapy
- B chemotherapy
- C chlamydia infection
- D cow's milk allergy
- E cytomegalovirus infection
- F gastro-oesophageal reflux
- G HIV infection
- H inhaled corticosteroid therapy
- I vitamin D deficiency
- J zinc deficiency

Questions 8, 9 and 10

- A asthma
- B costo-chondritis
- C epidemic myalgia
- D endocarditis
- E myocardial ischaemia
- F pleuritic chest pain
- G pneumothorax
- H precordial catch syndrome
- I psychogenic chest pain
- J reflux oesophagitis
- K slipping rib syndrome

Match the above diagnoses with the case histories given below:

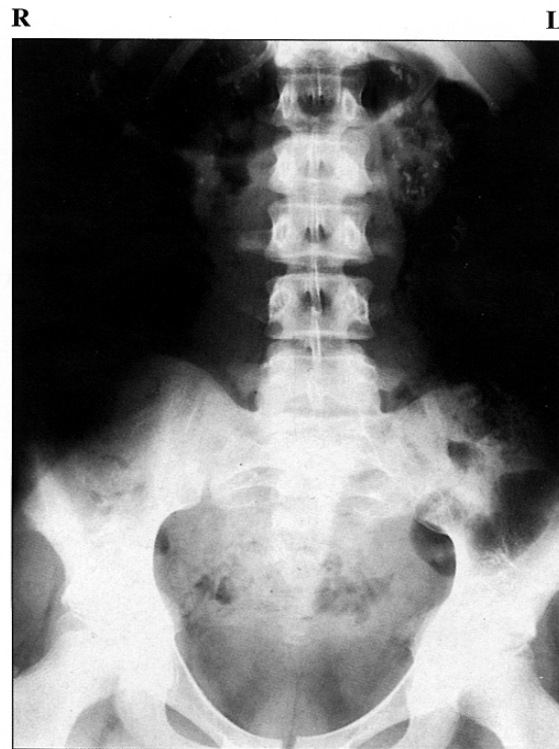
Note: *Each option may be used once, more than once or not at all.*

8. A boy of 12 presents with a 5 year history of chest pain occurring 2 or 3 times a month. The pain occurs at any time and is not particularly related to exertion. He describes the pain as a sharp stabbing pain localised to the left anterior chest wall which lasts for a few seconds. There are no associated symptoms although he is rather anxious with an increased heart rate after attacks of the pain. He has a normal exercise tolerance. Examination is normal. (3 marks)

This question continues on the next page.

9. A boy of 15 presents as an emergency with a 1 week history of chest pain. The pain first started when playing football. He also complained of headache and neck stiffness. The pain is localised to the left precordium and is described as aching and aggravated by exertion. He vomited twice on the day before presenting. On examination temperature 38.3°C, pulse 72, blood pressure 130/85mmHg. Mild neck stiffness. Heart sounds normal with no murmur. ECG sinus rhythm. ST elevation in the inferolateral leads. (3 marks)
10. A boy of 13 presents with a 3-month history of chest pain on exertion. He is a keen football player and has had to miss a couple of games particularly during cold weather because of this problem. The pain is described as a tight feeling over the lower chest which is difficult to localise. The pain is association with breathlessness. His symptoms gradually resolve after resting. Examination is entirely normal. (3 marks)

Question 11



Q11

This is an x-ray of the abdomen in a 12-year-old girl who attended a school for children with learning difficulties and complained of recurrent abdominal pain.

11. What abnormality can be seen on the plain abdominal film? (3 marks)

SELECT ONE ANSWER ONLY

- A bilateral renal calculi
- B bilateral sacro-ileitis
- C calcification of the pancreas
- D calcified para-aortic lymph nodes
- E constipation
- F megacolon
- G nephrocalcinosis

Questions 12, 13 and 14

A 13-year-old girl with known sickle cell disease was admitted with fever, cough and malaise. She had been admitted on several previous occasions for sickle cell crises with colicky right upper quadrant abdominal pain.

She is the only child of Nigerian parents, born in the UK, and living in London. Her father is an accountant, mother a housewife and they live in a small modern flat. She has just returned from a holiday in Nigeria.

On examination she was anxious and moderately distressed. She was pale and slightly jaundiced. Her temperature was 37.8°C, her pulse 110/minute and her respiratory rate 36/minute. There was dullness to percussion over the right upper chest anteriorly and posteriorly with bronchial breathing heard in the same area. The liver edge was palpable 2cm below the costal margin but the spleen was not palpable. There was a slight tenderness on palpation in the right hypochondrium. Her blood pressure was 110/75 mmHg. There were no other abnormal signs.

A specimen of urine contained excess urobilinogen, trace of blood and leucocytes, but no protein, glucose or bilirubin. The urine was normal on microscopy.

Investigations

Hb	6.2 g/dl
Reticulocytes	18%
WBC $12.5 \times 10^9/l$	
neutrophils	$8.5 \times 10^9/l$
lymphocytes	$3.5 \times 10^9/l$
monocytes	$0.25 \times 10^9/l$
eosinophils	$0.25 \times 10^9/l$
plasma bilirubin	120 $\mu\text{mol/l}$
plasma alkaline phosphatase	300 IU/l (normal range age 90-350)
plasma sodium	148 mmol/l
plasma urea	7.6 mmol/l
Chest x-ray	opacification of the right upper zone
Oxygen saturation	96% in air

This question continues on the next page.

12. What is the most likely cause of her current febrile illness? (4 marks)

SELECT ONE ANSWER ONLY

- A staphylococcal pneumonia
- B mycoplasma pneumonia
- C pneumococcal pneumonia
- D pneumocystis carinii pneumonia
- E sickle cell crisis

13. What is the most likely cause of the earlier episodes of her upper abdominal pain? (3 marks)

SELECT ONE ANSWER ONLY

- A biliary colic
- B chronic pancreatitis
- C hepatitis B infection
- D renal colic
- E sickle cell crisis

This question continues on the next page.

14. What would be the next most useful investigation in establishing the cause of her upper abdominal pain? (2 marks)

SELECT ONE ANSWER ONLY

- A abdominal CT scan
- B abdominal ultrasound
- C HIDA scan
- D hepatitis serology
- E plain x-ray of abdomen

Question 15



Q15

A child with known congenital heart disease presents with this lesion.

15. What is the underlying diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A deep venous thrombosis
- B Factor V Leiden deficiency
- C idiopathic thrombocytopenic purpura
- D meningococcal septicaemia
- E protein S deficiency
- F subacute bacterial endocarditis
- G systemic lupus erythematosus

Question 16

A 2-week-old male infant, previously well, presented with vomiting.

Investigations:

Hb	12.9 g/dl
WBC	$18.5 \times 10^9/l$
neutrophils	$10.0 \times 10^9/l$
lymphocytes	$7.8 \times 10^9/l$
monocytes	$0.7 \times 10^9/l$
platelets	$604 \times 10^9/l$

blood glucose	4.2 mmol/l
→ plasma sodium	123 mmol/l
→ plasma potassium	6.2 mmol/l
plasma urea	9.5 mmol/l

Blood gases:

pH	7.33 (plasma hydrogen ion concentration 47 nmol/l)
PaO ₂	6.7 kPa (50 mmHg)
PaCO ₂	4.5 kPa (34 mmHg)
plasma bicarbonate	17.3 mmol/l
Base deficit	-7.8 mmol/l

16. What is the most likely diagnosis? (5 marks)

SELECT ONE ANSWER ONLY

- A Bartter's syndrome
- B congenital adrenal hyperplasia (21-hydroxylase deficiency)
- C cystic fibrosis
- D posterior urethral valves
- E pyloric stenosis
- F renal tubular acidosis
- G syndrome of inappropriate anti-diuretic hormone secretion (SIADH)

Questions 17 and 18



Q17 & Q18

The dentist reported this incidental finding in a 16-year-old female.

17. What is the diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A aberrant parathyroid
- B cavernous haemangioma
- C cystic hygroma
- D lingual thyroid
- E lymphoma
- F mucus retention cyst
- G peri-tonsillar abscess
- H rhabdomyosarcoma

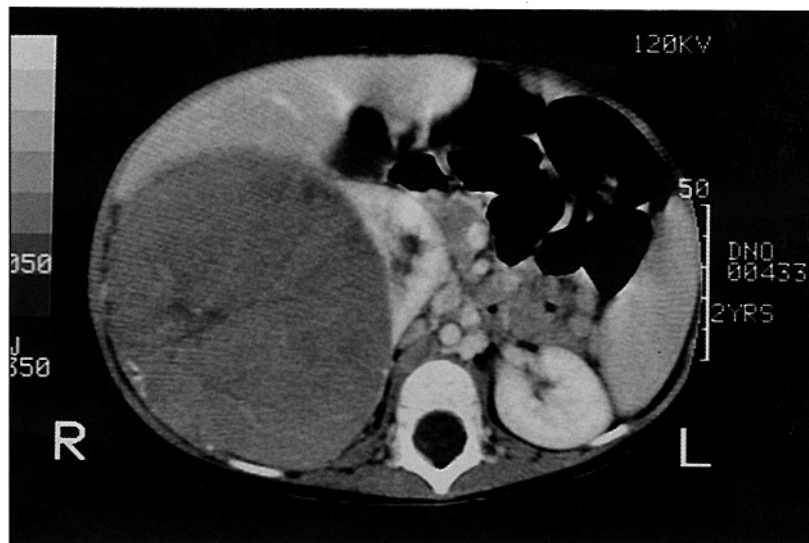
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18. How would you confirm the diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A chest x-ray
- B excision biopsy
- C MRI scan of head and neck
- D needle aspiration
- E parathyroid hormone levels
- F thyroid scan

Question 19 and 20



Q19 & Q20

A 4-year-old child presented with an abdominal mass.

19. What two abnormalities are seen on the CT scan with IV contrast? (4 marks)

SELECT TWO ANSWERS ONLY

- A displaced right kidney
- B inferior vena cava thrombosis
- C megacolon
- D multi-cystic right kidney
- E pancreatic pseudocyst
- F para-aortic lymph node enlargement
- G solid tumour of the liver
- H solid tumour of the right adrenal gland
- I solid tumour of the right kidney
- J spina bifida occulta

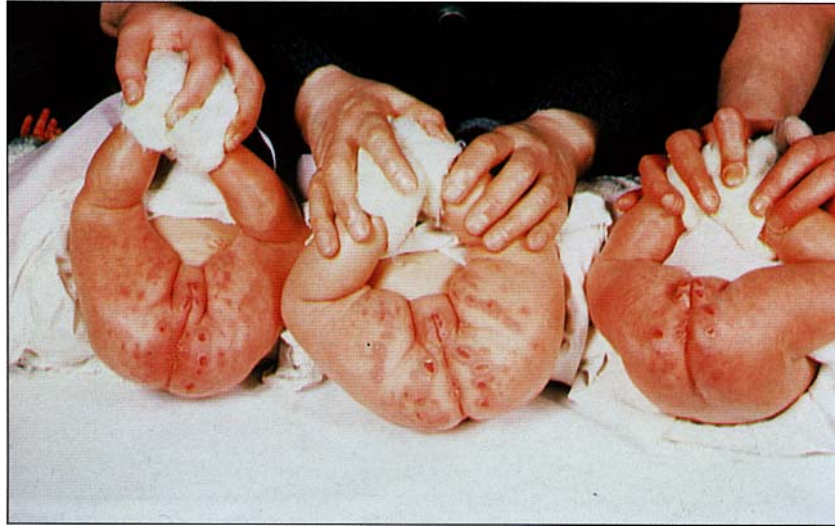
This question continues on the next page.

20. What is the likely diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A abdominal tuberculosis
- B hepatoma
- C Hirschprung's disease
- D lymphoma
- E neuroblastoma
- F pancreatic pseudocyst
- G polycystic renal disease
- H retroperitoneal abscess
- I Wilms' tumour

Question 21



Q21

21. What is the skin disorder from which these triplets are suffering? (3 marks)

SELECT ONE ANSWER ONLY

- A acrodermatitis enteropathica
- B ammoniacal dermatitis
- C eczema
- D herpes simplex infection
- E non-accidental injury
- F staphylococcal infection

Questions 22, 23 and 24

- A Addison's disease
- B aspirin toxicity
- C cerebral oedema
- D diabetic ketoacidosis
- E febrile convulsion
- F glycogen storage disease
- G hypopituitarism
- H medium chain acyl-coA dehydrogenase deficiency (MCAD)
- I Reye's syndrome

Match the following case descriptions with the most likely diagnosis from the list above.

Note: Each option may be used once, more than once, or not at all.

SELECT ONE ANSWER ONLY FOR EACH QUESTION

22. A 13-year-old girl has had poor diabetic control and has been feeling less well. She is admitted with diabetes ketoacidosis. Initial blood glucose is 27mmol/l with a pH of 7.0. She responds well to fluids, resuscitation and insulin and her blood glucose falls to 10 mmol/l. 12 hours after admission her level of consciousness deteriorates and her pulse drops to 70/min. (4 marks)
23. A 2-year-old boy becomes coryzal. He vomits three times and subsequently becomes drowsy followed by a generalised tonic clonic seizure. Blood glucose 4.6 mmol/l, sodium 137, potassium 4.5, creatinine 30, urea 2.5. (4 marks)
24. A 4-month-old male infant is admitted with a generalised seizure lasting ten minutes. The seizure is eventually terminated with rectal Diazepam. On examination he is noted to have a liver palpable five cm below the costal margin. Blood glucose 1.8 mmol/l. (4 marks)

Question 25



Q25

25. What is the cause of this lesion which had been present for three days? (3 marks)

SELECT ONE ANSWER ONLY

- A dacrocystitis
- B herpes simplex infection ← sparring the nose
- C herpes zoster infection ← if affecting the nose
- D human papillomavirus infection
- E molluscum contagiosum
- F staphylococcal infection
- G xanthoma

Question 26

A 10-year-old girl presented because of poor growth and four months of ill-defined abdominal pain. On examination her height was 128cm (10th centile) and weight 22kg (under 3rd centile). She had several mouth ulcers, appeared pale but examination was otherwise normal.

Preliminary investigation included:

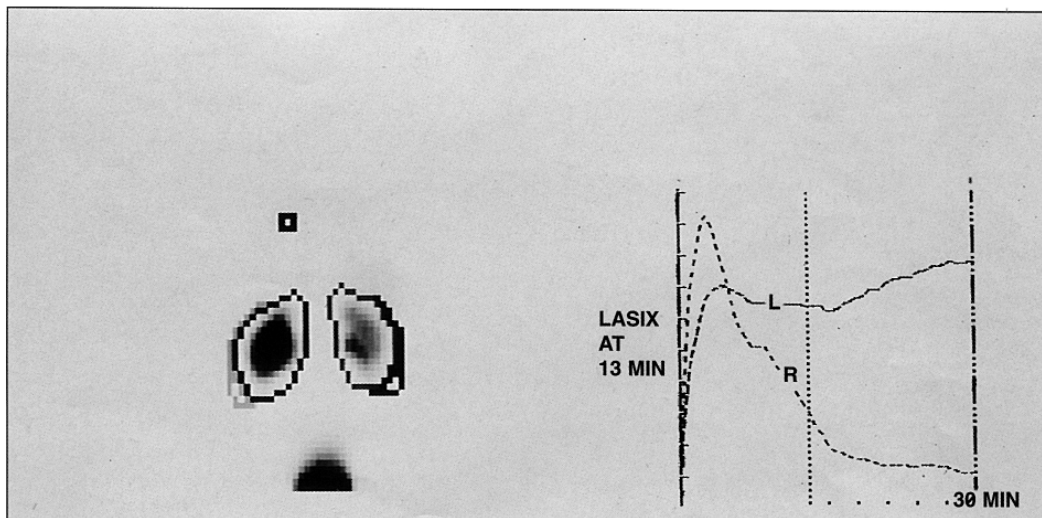
Hb	9.6 g/dl
MCH	26 pg
MCV	76 fl
WBC	$6.6 \times 10^9/l$
Platelets	$500 \times 10^9/l$
C-reactive protein	22 mg/l
Ferritin	15 µg/l (reference range 15-250µg/l)

26. What is the most likely cause of her problems? (5 marks)

SELECT ONE ANSWER ONLY

- A coeliac disease
- B Crohn's disease
- C eosinophilic colitis
- D iron deficiency anaemia
- E psychogenic pain
- F ulcerative colitis

Questions 27 and 28



Q27 & Q28

This is a MAG 3 renogram on a boy with recurrent urinary tract infection.

27. What is the important abnormality shown? (2 marks)

SELECT ONE ANSWER ONLY

- A bilateral pelvi-ureteric junction obstruction
- B bilateral renal cortical scarring
- C dilated left renal pelvis
- D dilated right renal pelvis
- E left pelvi-ureteric obstruction
- F left renal cortical scarring
- G right pelvi-ureteric obstruction
- H right renal cortical scarring
- I posterior urethral valves

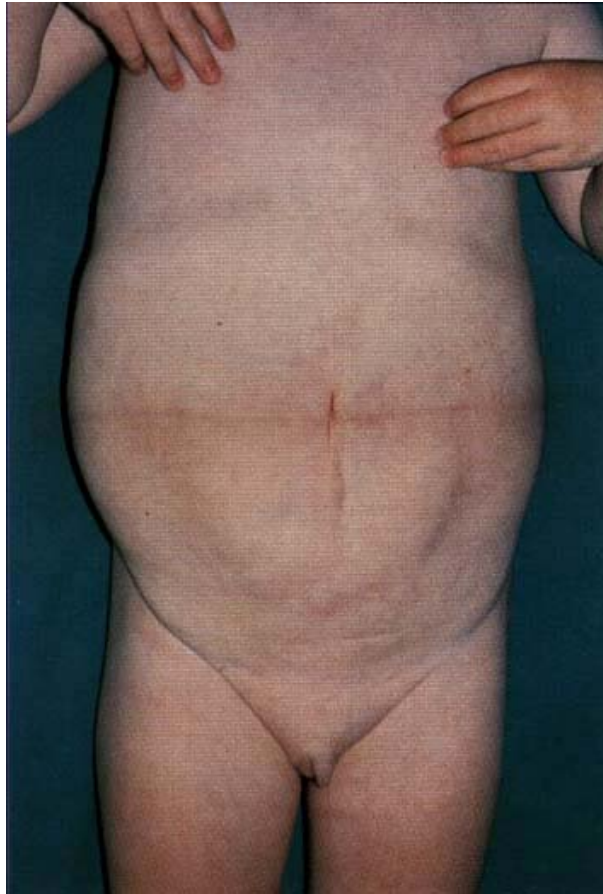
This question continues on the next page.

28. What abnormality is shown? (2 marks)

SELECT ONE ANSWER ONLY

- A bilaterally delayed excretion
- B bilateral ureteric reflux
- C delayed excretion on left despite furosemide
- D delayed excretion on right despite furosemide
- E left ureteric reflux
- F poor uptake on the left
- G poor uptake on the right
- H right ureteric reflux

Question 29



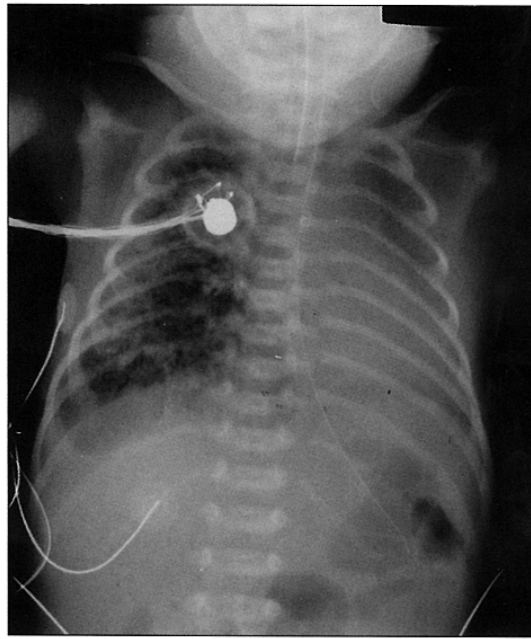
Q29

29. What are the two main abnormalities shown on this clinical photograph? (4 marks)

SELECT TWO ANSWERS ONLY

- A absent abdominal muscles
- B ambiguous genitalia
- C bladder distension
- D exomphalos
- E hypoplastic genitalia
- F scaphoid abdomen

Question 30



Q30

This is an x-ray of a 7-day-old baby with severe respiratory difficulty.

30. What is the most likely diagnosis of the abnormal right lung? (3 marks)

SELECT ONE ANSWER ONLY

- A bronchogenic cyst
- B congenital lobar emphysema
- C cystic adenomatoid malformation
- D diaphragmatic hernia
- E Macleod syndrome
- F meconium pneumonitis ← full term
- G pulmonary interstitial emphysema
- H pneumothorax

Question 31



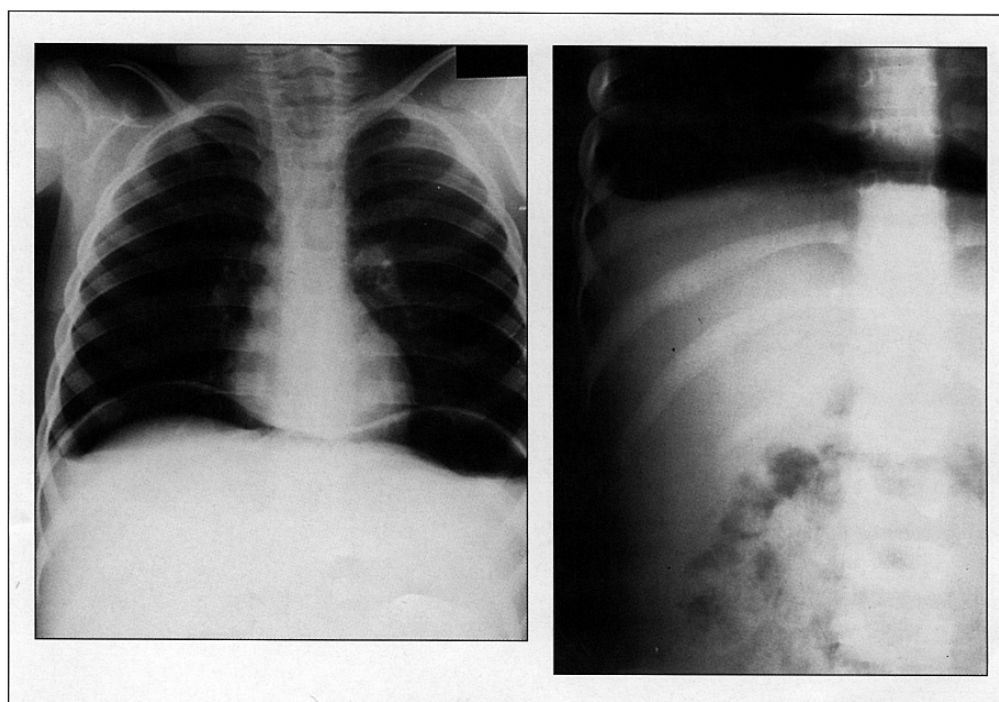
Q31

31. Select the three most likely causes for the appearance of this 4-year-old boy.

SELECT THREE ANSWERS ONLY

- A acute nephritis
- B bilateral ethmoid sinusitis
- C bilateral periorbital cellulitis
- D cardiac failure
- E cavernous sinus thrombosis
- F congenital angio-oedema
- G Henoch Schölein purpura
- H myotonia
- I nephritic syndrome
- J non-accidental injury
- K superior vena cava obstruction
- L water intoxication

Question 32



Q32

These x-rays were taken of an 18-month-old child on admission.

32. What two abnormalities do you see? (2 marks)

SELECT ONE ANSWER ONLY

- A ascites
- B bilateral sub-diaphragmatic air
- C fractured clavicle
- D hyperinflation
- E pneumomediastinum
- F pneumothorax
- G spina bifida occulta
- H subphrenic abscess

Question 33

A 13-year-old girl with poorly controlled insulin dependant diabetes presents to the clinic with a history of weight loss for three months.

She had developed diabetes at age 10 years when she presented with a history of weight loss, thirst and polyuria. A random blood glucose was 26 mmol/l and there was moderate ketonuria. Her serum contained islet-cell antibodies. She was started on insulin therapy using a pen device twice daily with mixed short and medium duration insulins. She entered a partial remission phase, during which her home glucose profiles were consistently in the range 4-10 mmol/l and her total daily insulin requirement fell to only 4 units before slowly increasing again.

During the last three months her control had started to deteriorate without obvious reason. Her total insulin dose was progressively increasing to over 60 units daily and she was changed on to a 4 injection per day regimen. Her record book showed few blood results but most were within the normal range. She complained of thirst and nocturia, mild intermittent abdominal pain and her weight had fallen from 45kg to 40kg.

She is the second of three daughters of a stable family with no financial worries. She had been successful and happy at school but recent events had caused school absences.

On examination she was tall, height 162cm (75-90 centile) and thin, weight 40kg (<25th centile). General examination was otherwise unremarkable. Her injection areas were not hypertrophied. She was at stage 2 puberty.

33. What is the most likely cause of her recent weight loss? (4 marks)

SELECT ONE ANSWER ONLY

- A anorexia nervosa
- B coeliac disease
- C hyperthyroidism
- D occult urinary tract infection
- E poor compliance with diet
- F poor compliance with insulin

THE END OF SPECIMEN PAPER 2

Paper Two Web Specimen Paper - Answer Key			
Question Number	Answer Key	Answer and Description	Marks
1	B	diabetes insipidus	3
2	G	response to DDAVP	3
3	D	mid-tracheal compression	2
	E	posterior oesophageal compression	2
4	H	vascular ring	2
5	A	fractures of radius and ulna	2
	I	rickets	2
6	D	oral candidiasis	1
7	B	chemotherapy	1
	G	HIV infection	1
	H	inhaled corticosteroid therapy	1
8	H	precordial catch syndrome	3
9	E	myocardial ischaemia	3
10	A	asthma	3
11	G	nephrocalcinosis	3
12	C	pneumococcal pneumonia	4
13	A	biliary colic	3
14	B	abdominal ultrasound	2
15	F	subacute bacterial endocarditis	3
16	B	congenital adrenal hyperplasia (21-hydroxylase deficiency)	5
17	D	lingual thyroid	2
18	F	thyroid scan	2
19	A	displaced right kidney	2
	I	solid tumour of the right kidney	2
20	I	Wilms' tumour	2
21	B	ammoniacal dermatitis	3
22	C	cerebral oedema	3
23	E	febrile convulsion	3
24	F	glycogen storage disease	3
25	B	herpes simplex infection	3
26	B	Crohn's disease	5
27	E	left pelvi-ureteric obstruction	2
28	C	delayed excretion on left despite furosemide	2
29	A	absent abdominal muscles	2
	F	scaphoid abdomen	2
30	G	pulmonary interstitial emphysema	3
31	A	acute nephritis	1
	F	congenital angio-oedema	1
	I	nephritic syndrome	1
32	B	bilateral sub-diaphragmatic air	2
33	F	poor compliance with insulin	4

CANDIDATE NUMBER:.....



Royal College of
Paediatrics and Child Health
Leading the way in Children's Health

Royal College of Paediatrics and Child Health

APPLIED KNOWLEDGE IN PRACTICE

PAPER 1

1. Complete the following:

Your full name (BLOCK LETTERS)

.....

RCPCH Number

Signature

2. Please check that your surname (family name) is correct on the **Answer Sheet**.
3. Using the pencil provided, answer all of the following 60 questions. **Do not** write anything on the Answer Sheet other than your response to each question.
4. Where questions are based upon photographs, x-rays, growth-charts, ECGs, or any other visual material, the question will refer you to the **Clinical Photographs Booklet**.
5. It is strictly forbidden to talk to, read the work of, or attempt in any way to communicate with, other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work. The College has tools which can identify copying of answers or collusion between candidates to share answers. In any situation the suspicion of guilt falls upon both parties until it can be proved otherwise. Breaches of these instructions, or misbehaviour in any other way, including continuing to write after the allotted time, may lead to suspension from the examination at the discretion of the invigilators. Serious breaches, such as cheating or colluding to gain advantage, could incur permanent suspension from College examinations.
6. Copyright law protects examination questions and the intellectual property of their authors. The unauthorised use of questions is a breach of copyright law.
7. Time allowed: 2 hours 30 minutes.

HOW TO COMPLETE THE ANSWER SHEET

The answer sheet contains a row of boxes for each question. Each box contains a letter. Indicate your answer(s) by filling in the appropriate box(es) with the pencil provided. The number of answers to be selected is stated in the question. If more than the stated number of answers are chosen, **no** marks will be awarded for the question. If you choose fewer answers than the stated number, marks will be given for each correct choice. The three types of question formats used in this examination are:

Best of List

In this question you are asked to choose the best single answer from a list.

Example 1

1. What is the most likely diagnosis?

SELECT ONE ANSWER ONLY

- A glucose 6 phosphate dehydrogenase deficiency.
- B haemophilia A.
- C haemorrhagic disease of the newborn.
- D hereditary spherocytosis.
- E Von Willebrand disease.

If you choose B this would be indicated on the answer sheet as:

1	<input type="radio"/> A	<input checked="" type="radio"/> B	<input type="radio"/> C	<input type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input type="radio"/> G	<input type="radio"/> H	<input type="radio"/> I	<input type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
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Choose n from many

In this question you are asked to choose a number of options from a longer list. For example the signs on a chest x-ray, or the findings on an ECG.

Example 2

2. What three investigations are most likely to be helpful in the next few hours?

SELECT THREE ANSWERS ONLY

- A abdominal ultrasound
- B blood culture
- C blood glucose
- D CRP
- E capillary blood gas analysis
- F chest x-ray
- G CT head scan
- H EEG
- I full blood count
- J lumbar puncture
- K plain abdominal x-ray
- L referral for surgical opinion
- M urea and electrolytes
- N urine culture

If you choose B, G and J this would be indicated on the answer sheet as:

2	<input type="radio"/> A	<input checked="" type="radio"/> B	<input type="radio"/> C	<input type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input checked="" type="radio"/> G	<input type="radio"/> H	<input type="radio"/> I	<input checked="" type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
.	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Extended Matching

In this question, a list of options is given at the beginning of the question. This might include a list of diagnoses, treatments, drugs or other management steps. After the list, short clinical scenarios or statements are made. You will be asked to choose the option(s) in the list which is(are) most appropriate.

Example 3

- A carbamazepine
- B clonazepam
- C ethosuximide
- D gabapentin
- E lamotrigine
- F phenytoin
- G prednisolone
- H topiramate
- I valproate
- J vigabatrin

Questions 3, 4 and 5

Choose the most appropriate drug from the list above in each of the following situations

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: Each answer may be used more than once

3. A 5 month old child presents to outpatients. Her mother tells you . . .
4. An 8 year old boy presents with episodes of collapse at school His teacher...
5. A 5 year old girl has a long history of epilepsy. She has previously been treated...

If you choose J, I and D respectively, this would be indicated on the answer sheet as:

3	<input type="radio"/> A	<input type="radio"/> B	<input type="radio"/> C	<input type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input type="radio"/> G	<input type="radio"/> H	<input type="radio"/> I	<input checked="" type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
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4	<input type="radio"/> A	<input type="radio"/> B	<input type="radio"/> C	<input type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input type="radio"/> G	<input type="radio"/> H	<input checked="" type="radio"/> I	<input type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
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General Instructions

1. Do not fold or crease the answer sheet or make any marks except in the spaces provided.
2. It is recommended that initially you indicate your answer against the questions in the question book. You should leave sufficient time (say half an hour) to transfer your answers to the answer sheet.
3. Answer the question by placing a neat bold horizontal line across the letter of your choice within the box.
4. The scoring machine has been programmed to ignore erasures (which can leave smudges) and this means that faint responses or partial responses may be ignored by the machine.
5. Smudges due to erasures may be caused by dirty rubbers – cleaning can be accomplished by rubbing against a clean part of the question book. Let the invigilator know if you are worried about the appearance of your erasures.

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Please do not turn over this page until instructed to do so by the invigilator.

Question 1

This 10 year old girl presented with a 2 day history of sore throat, noisy breathing, high fevers and difficulty swallowing.



1. Which of the following is the most likely causative organism for her symptoms?
(4 marks)

SELECT ONE ANSWER ONLY

- A Candida albicans
- B Cytomegalovirus
- C Epstein Barr virus
- D Haemophilus influenzae
- E Herpes simplex virus
- F Measles
- G Staphylococcus aureus

Question 2

This 6 month old boy has had a mild skin rash since the age of 2 months. He has suddenly become unwell with fever and marked worsening of the rash.

2. What is the most likely diagnosis? (3 marks)



SELECT ONE ANSWER ONLY

- A acrodermatitis enteropathica
- B chicken pox (varicella)
- C eczema herpeticum
- D impetigo
- E Langerhan's cell histiocytosis
- F primary Herpes simplex
- G Steven's Johnson syndrome
- H systemic candidiasis
- I toxic epidermal necrolysis

Questions 3, 4 and 5

The following is a list of cardiac diagnoses:

- A aortic regurgitation
- B aortic stenosis
- C atrial septal defect
- D coarctation of the aorta
- E mitral regurgitation
- F mitral stenosis
- G patent arterial duct (ductus arteriosus)
- H pulmonary stenosis
- I transposition of the great arteries
- J ventricular septal defect

For each of the following clinical scenarios select the most likely diagnosis:

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

3. A 2 year old boy presents with an asymptomatic murmur found on routine examination of the chest for an intercurrent chest infection. The peripheral pulses were normal but an ejection systolic murmur was heard over the right upper sternal area which radiated to the neck and along the left sternal edge. A systolic thrill was palpable in the suprasternal notch. (3 marks)
4. A 2 year old boy presents with an asymptomatic murmur found on routine examination of the chest for an intercurrent chest infection. There was an ejection systolic murmur and a thrill palpable in the left upper sternal area. The first and second heart sounds were clearly separated. (3 marks)
5. A 2 year old girl presents with an asymptomatic murmur found on routine examination of the chest for an intercurrent chest infection. There was a blowing systolic murmur best heard at the apex and radiating to the axilla. An apical low pitched systolic murmur was also heard. The second heart sound was increased but not split. (3 marks)

Question 6

An 8 year old boy presents to outpatients having recently noticed the development of pubic hair. He is having difficulty at school and is disruptive in class. He has a history of asthma for which he is taking inhaled fluticasone at the recommended dose for age. On examination there are no abnormalities detected apart from pubic hair, his testicular volumes are 2 mls bilaterally.

Investigations

Blood

sodium	128 mmol/l
potassium	5.8 mmol/l
bicarbonate	22 mmol/l
urea	3.4 mmol/l
creatinine	62 µmol/l

6. Which of the following is the most likely enzyme deficiency in his case? (4 marks)

SELECT ONE ANSWER ONLY

- A 11-beta hydroxylase
- B 17 alpha-hydroxylase
- C 21-hydroxylase
- D 3 alpha reductase
- E 3 beta-hydroxy-steroid dehydrogenase

Question 7

7. Which two of the following types of fractures would make you particularly suspicious of physical abuse? (4 marks)

SELECT TWO ANSWERS ONLY

- A complete fracture of the clavicle in a 3 year old
- B fracture of the femur in an ambulant child
- C fracture of the tibia in an ambulant child
- D posterior rib fracture under 1 year of age
- E spiral fracture of the humerus in a non-ambulant child
- F spiral fracture of the humerus in an ambulant child

Question 8

An 18 month old boy presented with a history of persistent cough for 4 weeks. He had an URTI 3 weeks previously, but had recovered with only symptomatic treatment. On examination his chest was clear to auscultation, his heart sounds were normal and his abdomen was soft with no organomegaly present. His chest x-ray is shown.



8. What is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A congenital diaphragmatic hernia
- B congenital lobar emphysema
- C congenital lung cyst
- D loculated pneumothorax
- E pneumatocele

Question 9

A 14 year old girl presented with a 3 week history of facial rash and similar rash over her shoulders following exposure to the sun. This was followed by increasing tiredness and malaise, swelling of the right thumb and the proximal interphalangeal joint of the right middle finger, shortness of breath on exertion and a dry cough. In the previous week she had complained of left sided pleuritic pain.

Her appearance is shown. Her temperature was 37.8°C. She had a tender spot in the pulp of both thumbs. She had a tachycardia, a pleuro-pericardial rub and crepitations at the left base.



Investigations

Blood

haemoglobin	10.5 g/dl
white cell count	$3.2 \times 10^9/l$
C-reactive protein	49 mg/l
blood culture	awaited

This question continues on the next page.

9. Which of the following investigations is most likely to lead to the diagnosis?
(4 marks)

SELECT ONE ANSWER ONLY

- A anti-double stranded DNA
- B Mycoplasma titres
- C Parvovirus titres
- D rheumatoid factor
- E skin swab
- F urinary porphyrin

Questions 10, 11 and 12

The following is a list of anti-inflammatory drugs:

- A anti-TNF alpha
- B azathioprine
- C ciclosporin
- D cyclophosphamide
- E intravenous immunoglobulin
- F oral prednisolone
- G methotrexate
- H rituximab
- I sulphasalazine
- J tacrolimus

Which of the above medications is most likely to be the cause of each of the following clinical scenarios?

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

- 10. A 7 year old boy with severe asthma and multiple hospital admissions has reduced growth velocity in the last 6 months. (3 marks)
- 11. A 16 year old girl with steroid resistant Crohn's disease develops neutropenia on alternative treatment. (3 marks)
- 12. A 5 year old boy 3 weeks post BMT for relapsed ALL develops macroscopic haematuria. (3marks)

Question 13

A 4 day old boy born to consanguineous Asian parents was brought to the emergency department with left sided focal fits. The fits stopped after giving rectal diazepam. On examination his temperature was 38°C, heart rate 140/minute and respiratory rate 40/minute.

He was drowsy post diazepam but rousable. The rest of the physical examination was normal.

13. Apart from a septic screen, including a lumbar puncture, which two of the following investigations would be most helpful in his immediate management? (4 marks)

SELECT TWO ANSWERS ONLY

- A ammonia
- B blood sugar
- C calcium
- D coagulation screen
- E cranial ultrasound
- F CT head scan
- G full blood count
- H urea and electrolytes

Question 14

A 14 month old boy has had one confirmed Coliform urinary tract infection. He was treated with a 7 day course of oral antibiotics and is now well.

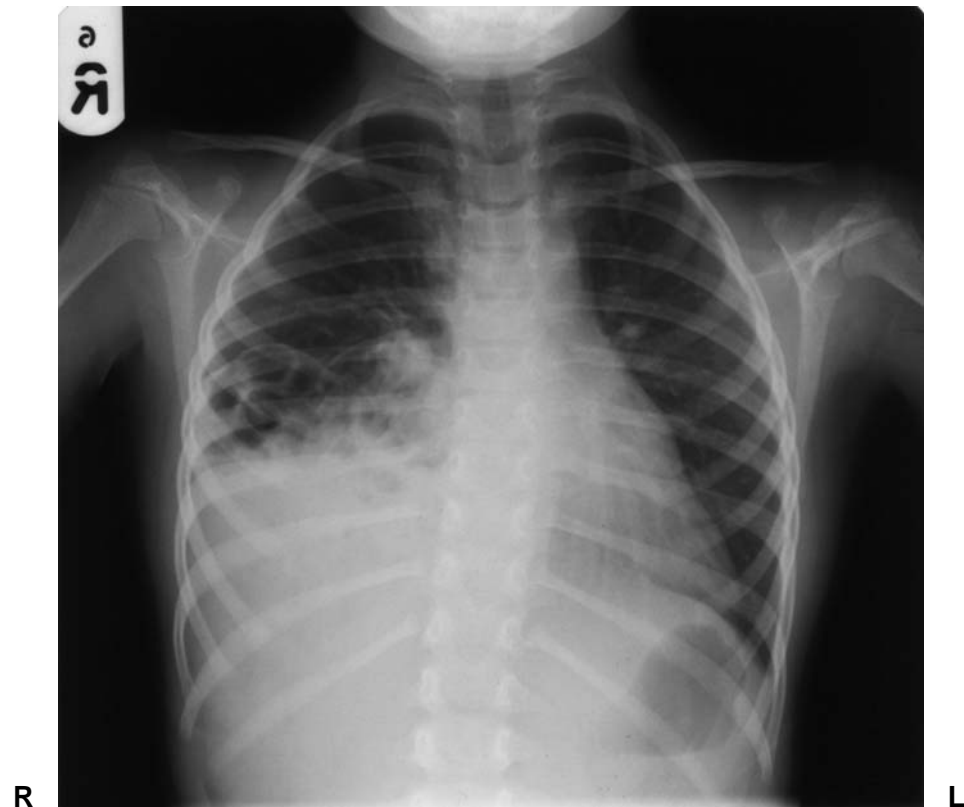
14. Which of the following investigations is most appropriate at this stage? (3 marks)

SELECT ONE ANSWER ONLY

- A DMSA scan
- B micturating cystogram
- C ultrasound scan of renal tract
- D all of the above
- E none of the above

Question 15

This is the chest x-ray of an 18 month old girl who presented with a history of cough, fever and noisy breathing for the previous 48 hours. She has previously been well and her height and weight were on the 25th to 50th centiles for age.



15. Which of the following is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A bronchiectasis
- B congenital cystic adenomatoid malformation
- C diaphragmatic hernia
- D foreign body aspiration
- E pneumatoceles
- F primary ciliary dyskinesia
- G pulmonary tuberculosis

Question 16

This infant was referred from the midwifery led unit having been born at term following an uneventful pregnancy. His clinical photograph is shown below.



16. What is the most important initial investigation? (4 marks)

SELECT ONE ANSWER ONLY

- A chromosome analysis
- B cranial CT scan
- C cranial ultrasound
- D EEG
- E full blood count
- F intraocular pressure measurement

Questions 17, 18 and 19

The following is a list of treatment options for seizures:

- A carbamazepine
- B lamotrigine
- C levetiracetam
- D no treatment
- E phenobarbitone
- F phenytoin
- G sodium valproate
- H topiramate
- I vigabatrin
- J zonisamide

Choose the most appropriate treatment for each of the following cases:

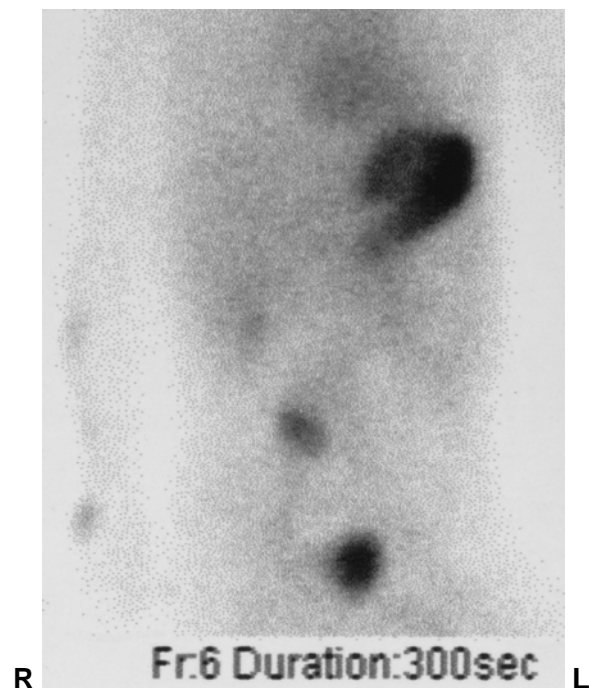
SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

- 17. A 13 year old girl with juvenile absence epilepsy. (3 marks)
- 18. A 6 month old boy with infantile spasms and developmental delay. (3 marks)
- 19. An 8 year old boy with infrequent nocturnal seizures and bilateral centrotemporal spikes on EEG. (3 marks)

Question 20

A 2 year old boy presented with a history of recurrent episodes of colicky abdominal pain for 2 months. His abdomen appeared bloated at times. His bowel motions were regular, but he had passed a blood stained stool on 3 separate occasions. His 99mTc pertechnetate scan is shown in the figure below.



20. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A appendix abscess
- B intussusception
- C malrotation
- D Meckel's diverticulum
- E vesico-urteric reflux

Question 21

A 15 year old boy was referred by his GP with a 48 hour history of painful swelling of the left side of his face. He was febrile, temperature 38.5⁰C, there was no significant cervical lymphadenopathy and no rash was present. He complained of severe pain when eating or drinking and had difficulty swallowing.

21. Which of the following is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A Ebstein-Barr virus infection
- B lymphoma
- C mumps
- D sarcoidosis
- E submandibular abscess

Question 22

A 4 month old baby boy from Eastern Europe who has recently immigrated to the UK presents with poor feeding, loose stools and faltering growth. On examination his length and weight are on the 2nd centiles for age. He is noted to have oral candidiasis and a mild degree of bilateral parotid swelling.

Investigations

Blood

haemoglobin	8.3 g/dl
white cell count	$5.2 \times 10^9/l$
neutrophils	$3.1 \times 10^9/l$
lymphocytes	$2.0 \times 10^9/l$
platelets	$485 \times 10^9/l$
immunoglobulins	IgG 9.5 g/dl (2.1-7.7)
	IgA 1.2 g/dl (0.05-0.4)
	IgM 1.8 g/dl (0.15-0.7)

Chest x-ray bilateral patchy infiltrates

22. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A chronic granulomatous disease
- B common variable immune deficiency
- C HIV/AIDs
- D severe combined immune deficiency
- E X-linked hypogammaglobulinaemia

Question 23

This 9 month old male infant presents with a 3 day history of fever, rhinorrhoea and rash. His mother had brought him to see the GP, who had prescribed erythromycin and antipyretics. The child did not improve and his rash worsened extending from his face to the rest of his body, in particular the axillae and groins.



23. What is the diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A atopic dermatitis
- B Herpes simplex infection with secondary bacterial infection
- C impetigo
- D Staphylococcal scalded skin syndrome
- E Stevens-Johnson syndrome
- F toxic epidermal necrolysis

Question 24

A 15 year old girl with focal epilepsy comes to clinic having had 5 seizures in the past 2 weeks. Her seizures were previously well controlled on carbamazepine. She was otherwise well except for a cough over the past month for which she has been taking erythromycin. She has recently started on the oral contraceptive pill.

24. What would be the next step with management? (3 marks)

SELECT ONE ANSWER ONLY

- A Add other anti-epileptic drug
- B Check carbamazepine level
- C Consider another form of contraception
- D CT head scan
- E Increase dose of carbamazepine

Question 25

This 9 month old infant was cyanosed at birth and had a cardiac operation at 3 months of age.



25. What condition is shown here? (3 marks)

SELECT ONE ANSWER ONLY

- A left sided Horner's syndrome
- B left sided lower motor neurone facial palsy
- C left sided ptosis
- D left sided upper motor neurone facial palsy
- E right-sided Horner's syndrome
- F right-sided lower motor neurone facial palsy
- G right-sided ptosis
- H right-sided upper motor neurone facial palsy

Question 26

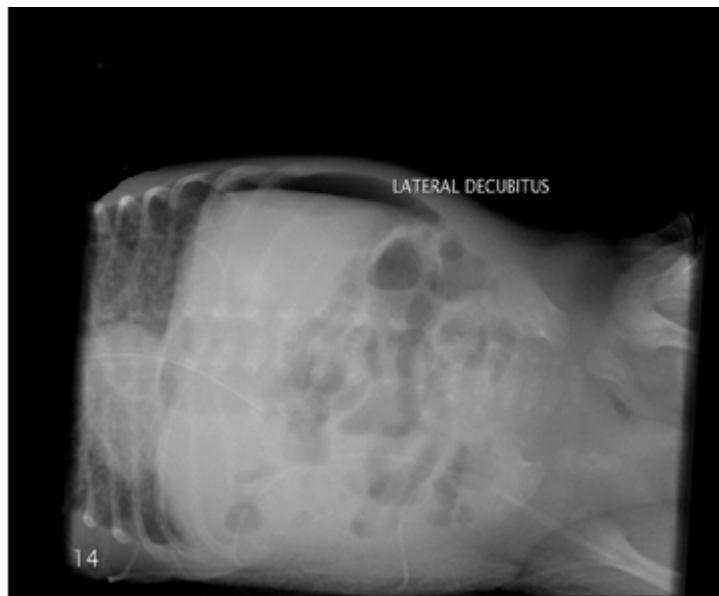
This 28 week gestation baby, currently ventilated, has had a worsening metabolic acidosis over the last 24 hours. His heart rate is 180/minute and mean blood pressure is 26 mmHg. He has developed peripheral oedema. Due to bilious aspirates an abdominal x-ray was performed.

Investigations

Blood gas

pH	7.01
pCO ₂	5.5kPa (41.2 mmHg)
pO ₂	8.1kPa (61 mmHg)
HCO ₃	10.2 mmol/l
BE	-17.0 mmol/l

This is his abdominal x-ray.



26. What is the most appropriate immediate management? (4 marks)

SELECT ONE ANSWER ONLY

- A administer 1mmol/kg sodium bicarbonate
- B fluid resuscitation with 10mls/kg 0.9% saline
- C increase ventilation pressures and rate
- D needle aspiration
- E start intravenous antibiotics

Question 27

A 12 year old boy with learning disability was seen in the child development centre with a 12 month history of increasingly awkward gait and difficulty getting footwear. On examination he has a scoliosis, marked pes cavus, absent lower limb reflexes and extensor plantar responses.

27. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A dermatomyositis
- B Duchenne muscular dystrophy
- C Friedreich's ataxia
- D hereditary motor and sensory neuropathy
- E spinal cord tumour

Question 28

A 12 year old boy was admitted with a 2 day history of fever, vomiting and diarrhoea. His parents were from Singapore. He had recently returned from a trip to Malaysia. Physical examination revealed a slightly lethargic looking boy with dry mucus membranes, but otherwise comfortable with good peripheral perfusion. Cardio-respiratory examination was unremarkable. The abdomen was not distended or tender; his spleen was palpable 3cm below the left costal margin. He was commenced on intravenous rehydration.

Investigations

Blood

haemoglobin	12.8 g/dl
MCV	56.8 fl
white cell count	$4.8 \times 10^9/l$
platelets	$210 \times 10^9/l$

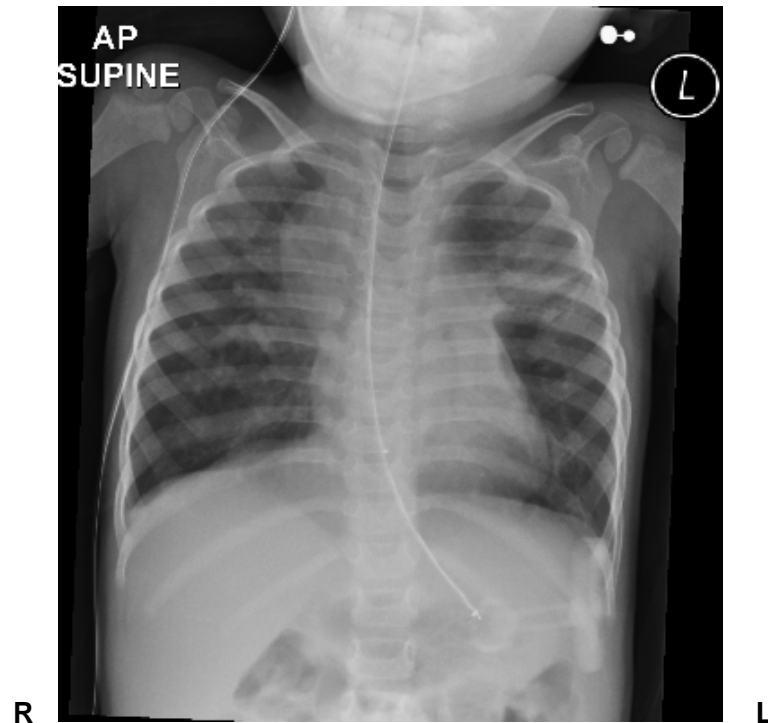
28. What is the most appropriate investigation to confirm the cause of the splenomegaly? (3 marks)

SELECT ONE ANSWER ONLY

- A blood film examination for malarial parasites
- B bone marrow aspirate
- C haemoglobin electrophoresis
- D glucose-6-phosphate dehydrogenase level
- E osmotic fragility test
- F serum ferritin

Questions 29 and 30

A 20 month old girl had a recurrent cough, intermittent vomiting and failure to thrive. She had been born at 29 weeks gestation and was oxygen dependent till 10 months of age. Her chest x-ray is shown in the figure below.



29. Which of the following investigations is most appropriate at this stage? (4 marks)

SELECT ONE ANSWER ONLY

- A bronchoscopy
- B CT scan thorax
- C echocardiography
- D serum immunoglobulins
- E sputum culture
- F video-swallow ← pH study

This question continues on the next page.

30. What treatment should this girl be receiving? (3 marks)

SELECT ONE ANSWER ONLY

- A antibiotics
- B anti-reflux therapy
- C anti-viral agents
- D chemotherapy
- E removal of thoracic mass

Question 31

This is the face of a child aged 3 months with a normal platelet count who is otherwise well.



31. What is the best description of this lesion? (3 marks)

SELECT ONE ANSWER ONLY

- A cavernous haemangioma
- B cystic hygroma
- C haemangioma
- D Kasabach-Meritt syndrome
- E mixed capillary/cavernous haemangioma
- F port wine naevus
- G strawberry naevus

Questions 32, 33 and 34

The following is a list of endocrine diagnoses:

- A constitutional growth and pubertal delay
- B growth hormone deficiency
- C hypothalamic hamartoma
- D idiopathic precocious puberty
- E Klinefelter syndrome
- F McCune – Albright syndrome
- G premature adrenarche
- H premature thelarche
- I testicular tumour
- J Turner syndrome

For each of the following clinical scenarios choose the most likely diagnosis.

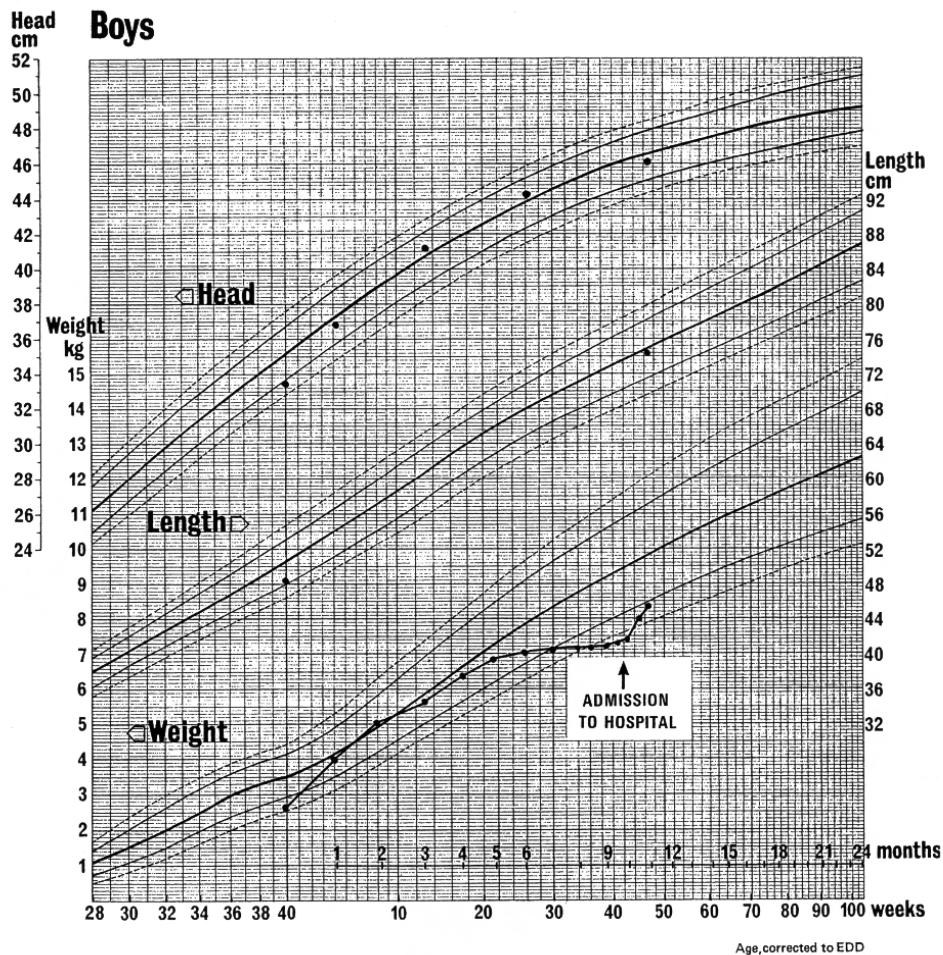
SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

- 32. A 2 year old girl with isolated bilateral breast development. (3 marks)
- 33. A 14 year old girl with short stature, delayed menarche and a cardiac murmur. (3 marks)
- 34. A 14 year old boy with short stature and a family history of delayed puberty. (3 marks)

Questions 35 and 36

This is the growth chart of a 10 month old boy who was admitted to hospital because of vomiting and poor weight gain. The baby fed well on a mixed diet. The stools were normal. There were no abnormal findings apart from wasting.



This question continues on the next page.

35. What is the most likely diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A coeliac disease
- B cow's milk protein intolerance
- C cystic fibrosis
- D gastro-oesophageal reflux disease
- E inadequate calorie intake

36. In the light of this growth chart, select the single most helpful next action.
(2 marks)

SELECT ONE ANSWER ONLY

- A discharge planning meeting
- B food diary for one week
- C measure IgA anti-tissue transglutaminase antibodies
- D oesophageal pH monitoring
- E RAST test for cow's milk IgE

Question 37

Objective: Is the use of intravenous immunoglobulin in newborn infants with isoimmune haemolytic jaundice effective in reducing the need for exchange transfusion?

Design: Systematic review of randomised and quasi-randomised controlled trials.

Method: Seven studies were identified. Three of these fulfilled the inclusion criteria and included a total of 189 infants, out of which two did not describe the method of allocation concealment. None of the studies used a placebo in the control group or described any method of blinding of intervention after allocation. Term and preterm infants with rhesus and ABO incompatibility were included.

Outcome: use of exchange transfusion.

Results: Relative risk of the use of exchange transfusion in the immunoglobulin treated group was 0.28, (95% confidence intervals (CI) 0.17 - 0.47) and numbers needed to treat (NNT) was 2.7. Weighted Mean Difference of exchange transfusions per infant in the immunoglobulin treated group was: -0.52 (95% CI -0.70 to -0.35).

Based on the information provided above, which of the following statements is true?
(3 marks)

SELECT ONE ANSWER ONLY

- A NNT of 2.7 means that 2.7 exchange transfusion would have been required for each intravenous immunoglobulin administration.
- B The number of exchange transfusions per infant was less in the group which did not receive immunoglobulin.
- C The result of this systematic review is influenced by the lack of use of placebo in the control group.
- D There was a reduction in the need for exchange transfusion in those treated with intravenous immunoglobulin.
- E This systematic review provides robust evidence for the routine use of intravenous immunoglobulin for the treatment of isoimmune haemolytic jaundice.

Question 38

A 3 day old baby girl was admitted with poor feeding, recurrent apneas and poor peripheral pulses. She required resuscitation and ventilatory support. Arterial blood gases showed a severe metabolic acidosis. She responded well to prostacycline infusion and oxygen saturation increased from 84% to 95%.

38. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A interrupted aortic arch
- B sepsis
- C transposition of great arteries ← no cyanosis and no vsd
- D tricuspid atresia
- E urea cycle disorder

Question 39

A 4 week old baby boy is seen in outpatients with a history of sudden jerky movements noticed by his mother almost from birth. They are seen most prominently whilst the baby is asleep. His mother thinks they are more obvious recently and may number from one or two to about ten or more in a few hours sleep. Each jerk seems to last a few seconds and appears to involve an isolated limb. Physical examination is normal.

39. Which of the following is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A benign sleep myoclonus
- B drug withdrawal syndrome
- C hypocalcaemia
- D infantile spasms
- E myoclonic epilepsy

Question 40

A 4 year old child presents to outpatients with his mother, father and elder brother. He has been referred with a pansystolic murmur at the apex heard by a GP at a pre-school medical. During the course of the examination he is noted to be on the 90th centile for height and the 25th centile for weight and has an asymmetrical chest. He has hypermobile joints and a history of joint pains.

40. Select the two most appropriate initial clinical responses to this scenario. (4 marks)

SELECT TWO ANSWERS ONLY

- A an orthopaedic opinion
- B ASO titre
- C chromosomal markers for Marfan's syndrome
- D echocardiography
- E enquire about a family history of sudden death
- F examine family members for similar characteristics

Question 41

Study: Parent initiated prednisolone for acute asthma in children of school age.

Objective: To study the effect of oral prednisolone on acute asthma symptoms in children aged 5-12 years.

Design: Double Blind Randomised Placebo controlled cross-over trial.

Outcome: Daytime asthma symptom score, night-time symptom score and school absenteeism.

Results: 230 children followed up over 3 years. 131 participants contributed, 308 episodes asthma requiring treatment. 155 episodes were treated with prednisolone, 153 with placebo. Mean daytime score with prednisolone treatment was 15% lower (95% confidence intervals (C.I.) 2-26%). Mean night-time score treated with prednisolone 16% lower (95% C.I. 0-30%). School absenteeism was reduced by a mean of 0.4 days (95% C.I. 0.0-0.8 days).

Adapted from BMJ 2010 vol 340, page 518-19)

Which of the following conclusions can be reached from the information presented above? (6 marks)

SELECT TWO ANSWERS ONLY

- A Acute asthma substantially affects school attendance.
- B Parents of all children with asthma should be given a supply of prednisolone.
- C Parents of children with asthma aged 5-12 years should not be given prednisolone to deliver at home.
- D Prednisolone does not significantly reduce night-time symptoms in asthma.
- E Prednisolone significantly reduces daytime symptom score in acute asthma.
- F The results are invalid as only 57% children received intervention

Question 42

An 18 month old girl is seen in her local community clinic. She is from a family who travel the country with her father who is a labourer. Her grandmother recently died of tuberculosis. The family were seen and the child's chest x-ray was normal. Four weeks previously she had been given BCG immunisation. She had had a cough and nasal discharge for 2 weeks. She was otherwise well and her temperature was 37.2⁰C. She has had no other immunisations. The health visitor notes a high non-attendance rate and points out the need to make progress in her immunisation programme.

Which of the following is the most appropriate course of action? (4 marks)

SELECT ONE ANSWER ONLY

- A Bring back in 1 week to give diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines.
- B Bring back in 6 weeks to give diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines. [deffer the MMR for a month so she will take all](#)
- C Offer diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines and measles, mumps and rubella immunisation immediately.
- D Offer diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines immediately.
- E Offer measles, mumps and rubella immunisation alone immediately.

Questions 43, 44 and 45

The following is a list of respiratory diagnoses:

- A bronchiectasis
- B Chlamydia pneumonitis
- C gastro-oesophageal reflux
- D hyper IgE syndrome
- E obliterative bronchiolitis
- F pertussis
- G primary ciliary dyskinesia
- H pulmonary tuberculosis
- I recurrent aspiration
- J tracheo-oesophageal fistula

For each of the following clinical scenarios choose the most likely diagnosis from the list above:

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

- 43. A 5 year old girl presents with a persistent cough following right lower lobe pneumonia aged 4. Crackles are heard in the right base. (3 marks)
- 44. A 2 year old boy who suffered from severe birth asphyxia presents with recurrent cough and wheeze. He had mild eczema as an infant. (3 marks)
- 45. A 3 year old boy with a history of recurrent skin sepsis, eczema, chronic cough and sputum production. Sputum samples grow Staphylococcus aureus. (3 marks)

Question 46

A baby boy, born at term, developed meconium aspiration syndrome requiring ventilation. His ventilator settings were PIP 28 cm H₂O, PEEP 5 cm H₂O, rate 50/minute, inspiratory time 0.4 seconds and FiO₂ 1.00. His heart rate was 140/minute and oxygen saturations 82%.

Blood gases

pH	7.21
pCO ₂	5.4 kPa (41 mmHg)
pO ₂	3.2 kPa (24 mmHg)
bicarbonate	18 mmol/l
base excess	-10 mmol/l

Which of the following is the appropriate change to make in management at this time?
(4 marks)

SELECT ONE ANSWER ONLY

- A add inhaled nitric oxide at 20ppm
- B half correct acidosis with a bicarbonate infusion
- C increase respiratory rate to 55/minute
- D start dobutamine at 5mcg/kg/hr
- E start high frequency oscillation (HFO)

hypoxia despite max ventilation
efforts — PPHTN

Question 47

A baby boy, birth weight 3.5 kg, is born at term in a district general hospital. The delivery involved a difficult forceps procedure following evidence of fetal distress and an abnormal CTG during labour. At birth there was no detectable heartbeat and the Apgar scores were 1 at 1 minute and 4 at 5 minutes. He was intubated and transferred to NICU.

Investigations

Blood gases 50 minutes post delivery

pH 7.0
pCO₂ 6.9 kPa (52 mmHg)
pO₂ 11.0 (82.5 mmHg)
bicarbonate 12.5 mmol/l
base excess -17 mmol

Pending transfer to the tertiary NICU, which of the following is the most appropriate action? (4 marks)

SELECT ONE ANSWER ONLY

- A give bicarbonate
- B give prophylactic phenobarbitone
- C hyperventilate the baby to reduce intracranial pressure
- D IV normal saline 10ml/kg bolus
- E passive cooling

Questions 48

A 7 year old boy was admitted with a 4 day history of multiple bruises all over his body. Four days prior to admission, he had a few small nose bleeds that subsided spontaneously. He was otherwise well; there was no mucosal bleeding and no organomegaly.

Investigations

Blood

haemoglobin	12.7 g/dl
white cell count	$7.2 \times 10^9/l$
neutrophils	$4.4 \times 10^9/l$
lymphocytes	$2.3 \times 10^9/l$
platelets	$3.0 \times 10^9/l$

Blood Film: Normal apart from thrombocytopenia

48. Which of the following is the most appropriate management? (3 marks)

SELECT ONE ANSWER ONLY

- A IV anit-D immunoglobulin
- B IV immunoglobulins
- C observation on the wards
- D oral prednisolone
- E platelet transfusion

Questions 49

A 10 year old girl, known to be asthmatic, is being treated with as required salbutamol and beclometasone 200mcg twice daily, presents with concerns about her growth. Four years previously she had been the tallest in her class and now she is the smallest. Her height is between the 25 and 50th centiles and weight on the 50 to 75th centile. She has a history of bloating over the past 2 years although her bowel habit has been normal.

Investigations

Blood

haemoglobin	12.5 g/dl
white cell count	10.9 x 10 ⁹ /l
normal differential	
platelets	235 x 10 ⁹ /l
ESR	20 mm/hr
CRP	30 mg/l
sodium	140 mmol/l
potassium	4 mmol/l
urea	4.8 mmol/l
creatinine	24 µmol/l

Which of the following investigations are the most likely to lead to a diagnosis? (4 marks)

SELECT TWO ANSWERS ONLY

- A abdominal ultrasound scan
- B IgA anti-tissue transglutaminase antibodies
- C IgF-1 level
- D Meckel's scan
- E short synacthen test
- F thyroid function tests

Question 50

A 15 year old boy with a diagnosis of asthma for 10 years was referred by his GP to outpatients because of persisting respiratory symptoms. At the time of referral he was on inhaled beclometasone 1 milligram twice daily and inhaled salmeterol 50 micrograms twice daily for the last 6 months. He was also on inhaled salbutamol 2-10 puffs as required. His Peak Expiratory Flow rate was 255 L/minute (71% predicted). He was not able to participate fully in school sports.

50. Which two of the following investigations would you do next? (4 marks)

SELECT TWO ANSWERS ONLY

- A bronchial lavage
- B bronchoscopy
- C chest x-ray
- D Mantoux test
- E nasopharyngeal brushings
- F short synacthen test
- G spiral CT of chest
- H sweat test
- I ventilation/perfusion lung scan

Question 51

A previously well 16 year old boy presented to outpatients with a 6 month history of frequent headaches. Recent problems with balance had occurred while playing football for his school team. The headaches were relieved by lying flat. There was a strong family history of migraine.

Physical examination was normal apart from a positive Romberg sign and unsteadiness on attempting to heel-toe walk. Reflexes were normal.

51. Which of the following is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

[schwa](#)

- A Arnold-Chiari malformation type 1
- B idiopathic (benign) intracranial hypertension
- C late onset muscular dystrophy (Becker)
- D migraine
- E spinal cord tumour

Question 52

A 12 year old boy with type 1 diabetes since the age of 8 comes to clinic for his annual review. His growth is normal for age, his control has been excellent and his HbA1c is 42 mmol/mol (6%).

52. Which of the following conditions known to be associated with type 1 diabetes should be screened for at this age? (6 marks)

SELECT TWO ANSWERS ONLY

- A coeliac disease
- B exocrine pancreatic insufficiency
- C hypercholesterolaemia
- D nephropathy
- E peripheral neuropathy
- F retinopathy

Question 53

A 12 year old girl presented to the outpatient department with a 3 month history of headache, morning vomiting and weight loss. Her mother had died in a road traffic accident 1 year previously. She had been attending a psychiatrist for symptoms of depression since that time.

On examination she was thin and mildly dehydrated. Apart from apparent difficulty with bilateral upward gaze her neurological examination was normal with no abnormality of limb power, tone or reflexes. There was no papilloedema and her blood pressure was 110/65 mmHg.

53. Which of the following is the most likely finding on the cranial MRI? (4 marks)

SELECT ONE ANSWER ONLY

- A herniation of the medulla
- B idiopathic (benign) intracranial hypertension
- C intracranial tumour
- D mid-brain haemorrhage causing superior oblique muscle paralysis (Parinaud syndrome)
- E normal scan
- F subarachnoid haemorrhage

Question 54

A 4 year old child presents with his 5th febrile seizure. He is developmentally normal and his father had a history of febrile seizures as a child.

54. Which of the following is most appropriate at this stage? (2 marks)

SELECT ONE ANSWER ONLY

- A CT head scan
- B ECG
- C EEG
- D MRI brain scan
- E no investigation

Question 55

A 10 month old boy presents to the emergency department at 7am after being found at home unrousable in his cot. Nursing staff note a few bruises over his forehead and limbs. He was born at term and spent 3 weeks on the neonatal unit with neonatal abstinence syndrome. His mother reports him to have been hot the previous night. On examination he is breathing comfortably and his oxygen saturations are 95% in room air. His Glasgow Coma Scale score is 13, his capillary refill time is 3-4 seconds.

55. What is the most appropriate initial investigation? (3 marks)

SELECT ONE ANSWER ONLY

- A blood gas
- B blood glucose
- C fundoscopy
- D lumbar puncture
- E urgent CT head scan

Question 56

A 2 year old African-Caribbean girl was referred with concern about her locomotor development. Further history taking revealed that she was largely breast fed and was described as a “fussy eater”. She lived with her mother and new boyfriend in a high rise council block. Her height and weight were on the 2nd to 9th centiles for age.

Her wrist x-ray is shown.



56. Which of the following is the most likely explanation for her symptoms? (3 marks)

SELECT ONE ANSWER ONLY

- A metaphyseal dysplasia
- B mucopolysaccharidosis
- C non-accidental injury
- D osteogenesis imperfecta
- E rickets
- F sickle cell anaemia

Question 57

A 9 month old boy presented to the emergency department with a 24 hour history of irritability and decreased movements of his left arm. His parents said that he had accidentally rolled off the couch onto the floor the previous evening. His x-ray is shown in figure below.



57. What is the most likely cause of this appearance? (3 marks)

SELECT ONE ANSWER ONLY

- A accidental injury
- B non-accidental injury
- C osteogenesis imperfecta
- D osteopenia
- E rickets

Question 58

A 12 year old girl with frontal lobe epilepsy presents with a 4 day history of fever cough vomiting and drowsiness.

Investigations

Blood

sodium	116 mmol/l
potassium	2.8 mmol/l
chloride	90 mmol/l
urea	1.8 mmol/l
creatinine	40 umol/l
bicarbonate	19 mmol/l
osmolality	247 mmol/kg
urine	107 mmol/kg
sodium	18 mmol/l
potassium	19 mmol/l
chloride	< 15 mmol/l

58. Which of the following is the most likely cause of her current biochemical findings?
(3 marks)

SELECT ONE ANSWER ONLY

- A Addison's disease
- B cerebral salt wasting
- C excessive water drinking
- D pseudohyponatremia
- E syndrome of inappropriate anti-diuretic hormone secretion (SIADH)

Question 59 and 60

A 7 year old boy presents with fever, cough, dyspnoea and generalized oedema. His chest x-ray is shown in the figure below.



59. What is the most appropriate investigation to establish the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A blood culture
- B CT thorax
- C echocardiography
- D Mycoplasma serology
- E pleural tap for culture
- F sputum culture

This question continues on the next page.

60. What is the most appropriate treatment? (3 marks)

SELECT ONE ANSWER ONLY

- A amoxicillin
- B chest tube drainage
- C erythromycin
- D furosemide
- E inhaled salbutamol
- F oral steroid

End of specimen paper

Royal College of Paediatrics and Child Health

APPLIED KNOWLEDGE IN PRACTICE

PAPER 1

Q No	No of Ans	Ans	Marks	Answer in words
1	1	C	4	Epstein Barr virus
2	1	C	3	eczema herpeticum
3	1	B	3	aortic stenosis
4	1	C	3	atrial septal defect
5	1	E	3	mitral regurgitation
6	1	C	4	21-hydroxylase
7	2	D	2	posterior rib fracture under one year of age
		E	2	spiral fracture of the humerus in a non-ambulant child
8	1	C	4	congenital lung cyst
9	1	A	4	Anti-double stranded DNA
10	1	F	3	oral prednisolone
11	1	B	3	azathioprine
12	1	D	3	cyclophosphamide
13	2	B	2	blood sugar
		C	2	calcium
14	1	E	3	none of the above
15	1	C	4	diaphragmatic hernia
16	1	F	4	intraocular pressure measurement
17	1	B	3	lamotrigine
18	1	I	3	vigabatrin
19	1	D	3	no treatment
20	1	D	3	Meckel's diverticulum
21	1	C	3	mumps
22	1	C	3	HIV/AIDS
23	1	D	4	Staphylococcal scalded skin syndrome
24	1	C	3	Consider another form of contraception
25	1	A	3	left sided Horner's syndrome
26	1	B	4	fluid resuscitation with 10mls/kg 0.9% saline
27	1	C	3	Friedreich's ataxia
28	1	C	3	haemoglobin electrophoresis
29	1	F	4	video-swallow
30	1	B	3	anti-reflux therapy
31	1	A	3	cavernous haemangioma
32	1	H	3	premature thelarche
33	1	J	3	Turner Syndrome
34	1	A	3	Constitutional growth and pubertal delay
35	1	E	2	inadequate calorie intake
36	1	A	2	discharge planning meeting
37	1	D	3	those treated with intravenous immunoglobulin
38	1	A	3	interrupted aortic arch
39	1	A	3	benign sleep myoclonus
40	2	D	2	echocardiography
		F	2	examine family members for similar characteristics
41	2	E	3	Prednisolone significantly reduces daytime symptom score in acute asthma
		E	3	Prednisolone does not significantly reduce night-time symptoms in asthma

Royal College of Paediatrics and Child Health

APPLIED KNOWLEDGE IN PRACTICE

PAPER 1

Q No	No of Ans	Ans	Marks	Answer in words
42	1	C	4	Offer diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines and measles, mumps and rubella immunisation
43	1	A	3	bronchiectasis
44	1	I	3	recurrent aspiration
45	1	D	3	hyper IgE syndrome
46	1	A	3	add inhaled nitric oxide at 20ppm
47	1	E	4	passive cooling
48	1	C	4	observation on the wards
49	1	B	2	IgA anti-tissue transglutaminase antibodies
		E	2	short synacthen test
50	2	C	2	chest x-ray
		F	2	short synacthen test
51	1	A	4	Arnold-Chiari malformation type 1
52	2	D	3	nephropathy
		F	3	retinopathy
53	1	C	4	intracranial tumour
54	1	E	2	No investigation
55	1	B	3	blood glucose
56	1	E	3	rickets
57	1	B	3	non-accidental injury
58	1	C	3	excessive water drinking
59	1	C	3	echocardiography
60	1	D	3	furosemide
THE END				

CANDIDATE NUMBER:.....



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APPLIED KNOWLEDGE IN PRACTICE

PAPER 2

1. Complete the following:

Your full name (BLOCK LETTERS)

.....

RCPCH Number

Signature

2. Please check that your surname (family name) is correct on the **Answer Sheet**.
3. Using the pencil provided, answer all of the following 60 questions. **Do not** write anything on the Answer Sheet other than your response to each question.
4. Where questions are based upon photographs, x-rays, growth-charts, ECGs, or any other visual material, the question will refer you to the **Clinical Photographs Booklet**.
5. It is strictly forbidden to talk to, read the work of, or attempt in any way to communicate with, other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work. The College has tools which can identify copying of answers or collusion between candidates to share answers. In any situation the suspicion of guilt falls upon both parties until it can be proved otherwise. Breaches of these instructions, or misbehaviour in any other way, including continuing to write after the allotted time, may lead to suspension from the examination at the discretion of the invigilators. Serious breaches, such as cheating or colluding to gain advantage, could incur permanent suspension from College examinations.
6. Copyright law protects examination questions and the intellectual property of their authors. The unauthorised use of questions is a breach of copyright law.
7. Time allowed: 2 hours 30 minutes.

HOW TO COMPLETE THE ANSWER SHEET

The answer sheet contains a row of boxes for each question. Each box contains a letter. Indicate your answer(s) by filling in the appropriate box(es) with the pencil provided. The number of answers to be selected is stated in the question. If more than the stated number of answers are chosen, **no** marks will be awarded for the question. If you choose fewer answers than the stated number, marks will be given for each correct choice. The three types of question formats used in this examination are:

Best of List

In this question you are asked to choose the best single answer from a list.

Example 1

1. What is the most likely diagnosis?

SELECT ONE ANSWER ONLY

- A glucose 6 phosphate dehydrogenase deficiency.
- B haemophilia A.
- C haemorrhagic disease of the newborn.
- D hereditary spherocytosis.
- E Von Willebrand disease.

If you choose B this would be indicated on the answer sheet as:

1	<input type="radio"/> A	<input checked="" type="radio"/> B	<input type="radio"/> C	<input type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input type="radio"/> G	<input type="radio"/> H	<input type="radio"/> I	<input type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
.	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

Choose n from many

In this question you are asked to choose a number of options from a longer list. For example the signs on a chest x-ray, or the findings on an ECG.

Example 2

2. What three investigations are most likely to be helpful in the next few hours?

SELECT THREE ANSWERS ONLY

- A abdominal ultrasound
- B blood culture
- C blood glucose
- D CRP
- E capillary blood gas analysis
- F chest x-ray
- G CT head scan
- H EEG
- I full blood count
- J lumbar puncture
- K plain abdominal x-ray
- L referral for surgical opinion
- M urea and electrolytes
- N urine culture

If you choose B, G and J this would be indicated on the answer sheet as:

2	⊂A	⊂B	⊂C	⊂D	⊂E	⊂F	⊂G	⊂H	⊂I	⊂J	⊂K	⊂L	⊂M	⊂N	⊂O	⊂P
.	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃

Extended Matching

In this question, a list of options is given at the beginning of the question. This might include a list of diagnoses, treatments, drugs or other management steps. After the list, short clinical scenarios or statements are made. You will be asked to choose the option(s) in the list which is(are) most appropriate.

Example 3

- A carbamazepine
- B clonazepam
- C ethosuximide
- D gabapentin
- E lamotrigine
- F phenytoin
- G prednisolone
- H topiramate
- I valproate
- J vigabatrin

Questions 3, 4 and 5

Choose the most appropriate drug from the list above in each of the following situations

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: Each answer may be used more than once

3. A 5 month old child presents to outpatients. Her mother tells you . . .
4. An 8 year old boy presents with episodes of collapse at school His teacher...
5. A 5 year old girl has a long history of epilepsy. She has previously been treated...

If you choose J, I and D respectively, this would be indicated on the answer sheet as:

3	⊂A	⊂B	⊂C	⊂D	⊂E	⊂F	⊂G	⊂H	⊂I	⊂J	⊂K	⊂L	⊂M	⊂N	⊂O	⊂P
.	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃
4	⊂A	⊂B	⊂C	⊂D	⊂E	⊂F	⊂G	⊂H	⊂I	⊂J	⊂K	⊂L	⊂M	⊂N	⊂O	⊂P
.	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃
5	⊂A	⊂B	⊂C	⊂D	⊂E	⊂F	⊂G	⊂H	⊂I	⊂J	⊂K	⊂L	⊂M	⊂N	⊂O	⊂P
.	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃	⊃

General Instructions

1. Do not fold or crease the answer sheet or make any marks except in the spaces provided.
2. It is recommended that initially you indicate your answer against the questions in the question book. You should leave sufficient time (say half an hour) to transfer your answers to the answer sheet.
3. Answer the question by placing a neat bold horizontal line across the letter of your choice within the box.
4. The scoring machine has been programmed to ignore erasures (which can leave smudges) and this means that faint responses or partial responses may be ignored by the machine.
5. Smudges due to erasures may be caused by dirty rubbers – cleaning can be accomplished by rubbing against a clean part of the question book. Let the invigilator know if you are worried about the appearance of your erasures.

BLANK PAGE

Please do not turn over this page until instructed to do so by the invigilator.

Question 1

The illustration shows the tongue of an otherwise healthy child. Her mother was worried about this appearance.



1. Which of the following is the most appropriate management? (3 marks)

SELECT ONE ANSWER ONLY

- A biopsy of the white area
- B determine HIV status
- C follow up for observation
- D nystatin
- E offer reassurance

Questions 2, 3 and 4

The following is a list of respiratory diagnoses:

- A aspergillosis
- B asthma
- C bronchiectasis
- D cystic fibrosis
- E foreign body aspiration
- F habitual cough
- G immune deficiency
- H pertussis
- I primary ciliary dyskinesia
- J tracheobronchomalacia

Choose the most likely diagnosis for each of the following scenarios:

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

2. A previously well 8 year old boy presents with a 2 month history of cough. He has episodes of nocturnal and daytime paroxysmal cough intermittently productive of clear sputum. He has had a trial of inhaled corticosteroids which have not had any benefit. He has been fully vaccinated for his age. (3 marks)
3. A 6 year old boy has a 6 month history of a loud, non-productive cough noticed by his mother at breakfast time. He has poor school attendance. His height and weight are on the 50th centile. (3 marks)
4. A 2 year old Caucasian boy with a history of chronic rhinitis and recurrent otitis media presents with a 4 week history of recurrent cough and purulent sputum production. (3 marks)

Question 5

A 3 day old girl presented with swelling and bruising of the right eyelid. She had been born at term by ventouse delivery due to fetal distress. Birth weight was 2.3 kg and she did not require resuscitation. She vomited 15 minutes after receiving oral vitamin K. She was otherwise well and feeding had been established without difficulty.

Investigations

Blood

haemoglobin	14.4 g/dl
white cell count	$6.5 \times 10^9/l$
normal differential	
platelets	$29 \times 10^9/l$
CRP	5 mg/l

5. Which of the following is the most important information contributing to the explanation of her problem? (4 marks)

SELECT ONE ANSWER ONLY

- A family history of easy bruising
- B low maternal platelet count in pregnancy
- C maternal anti-epileptic medication during pregnancy
- D perinatal risk factors for Group B Streptococcal sepsis
- E vomiting after vitamin K administration

Question 6

A 14 year old boy was referred to outpatients had gynaecomastia. He reported that his breast tissue had been there for the past year and that he has been embarrassed by it. There was no nipple pain or discharge. On examination his body mass index was 26 kg/m^2 . Testicular volume was 10 ml bilaterally. Pubic hair was at Tanner Stage 3.

6. What is the next most appropriate action? (3 marks)

SELECT ONE ANSWER ONLY

- A karyotype
- B MRI brain scan
- C perform GnRH stimulation test
- D reassure child and review him in 6 months time
- E serum LH and FSH
- F serum oestradiol
- G serum prolactin

Question 7

Title: Short versus long duration of antibiotic therapy for bacterial meningitis: a meta-analysis of randomised controlled trials in children.

Objective: To evaluate the effectiveness of short course (<7 days) versus long course (7-14 days) of antibiotic therapy for bacterial meningitis.

Outcomes: Clinical success, mortality, duration of hospitalisation, adverse events, hearing impairment, long-term neurological complications.

Results: Five open-label RCTs involving a total of 367 patients were included. The odds ratios for short versus long course outcomes were as follows: long-term neurological complications 0.60 (95% confidence interval (CI) 0.29-1.27), hearing impairment 0.59 (95% CI 0.28-1.23). The weighted mean difference in the duration of hospitalisation was -2.17 days (95% CI -3.85 to -0.50).

Adapted from ADC 2009;94:607-614

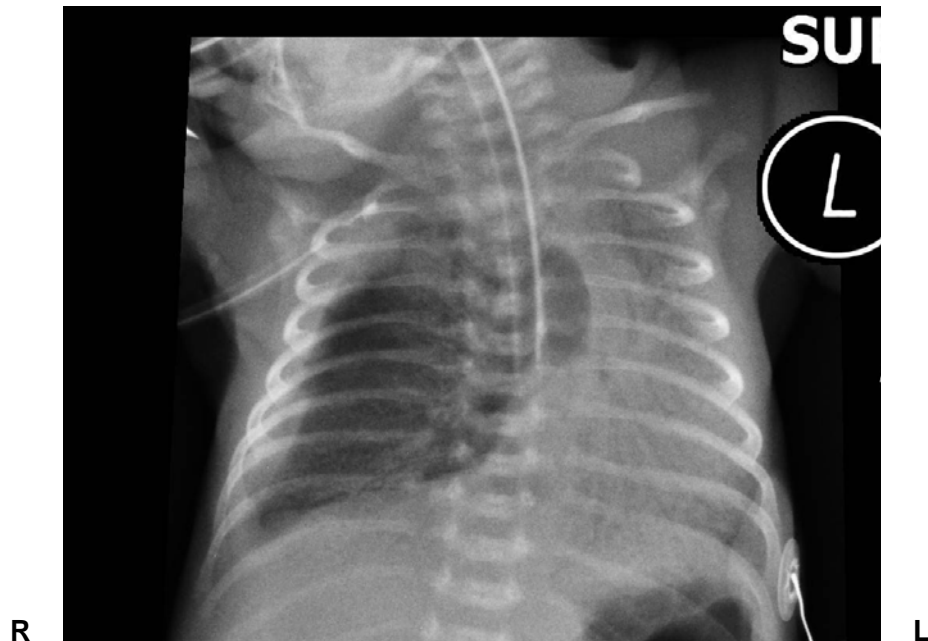
7. Which of the following conclusions can be deduced from the data? (4 marks)

SELECT TWO ANSWERS ONLY

- A Long course treatment resulted in higher levels of hearing impairment.
- B Meta-analysis of RCTs is not the most appropriate way to resolve such clinical issues.
- C Patients treated for meningitis should be observed in hospital for 24 hours following cessation of treatment.
- D Short course treatment resulted in a significantly reduced length of hospital stay.
- E Short course treatment should become the standard treatment for bacterial meningitis in children.
- F There was no difference in the level of neurological complications between treatment groups.

Question 8

A baby girl was born at 30 weeks gestation. She received Curosurf, was ventilated for 12 hours and then weaned to CPAP. On day 3 she suddenly deteriorated with desaturations and bradycardias. Chest x-ray showed a right pneumothorax and a chest drain was inserted. She was reintubated and ventilated on SIMV at pressures of 20/4 cmH₂O, rate 35/minute, I time 0.4s, in 27% oxygen. One hour later her condition worsened significantly. Her chest x-ray is shown in the figure below.



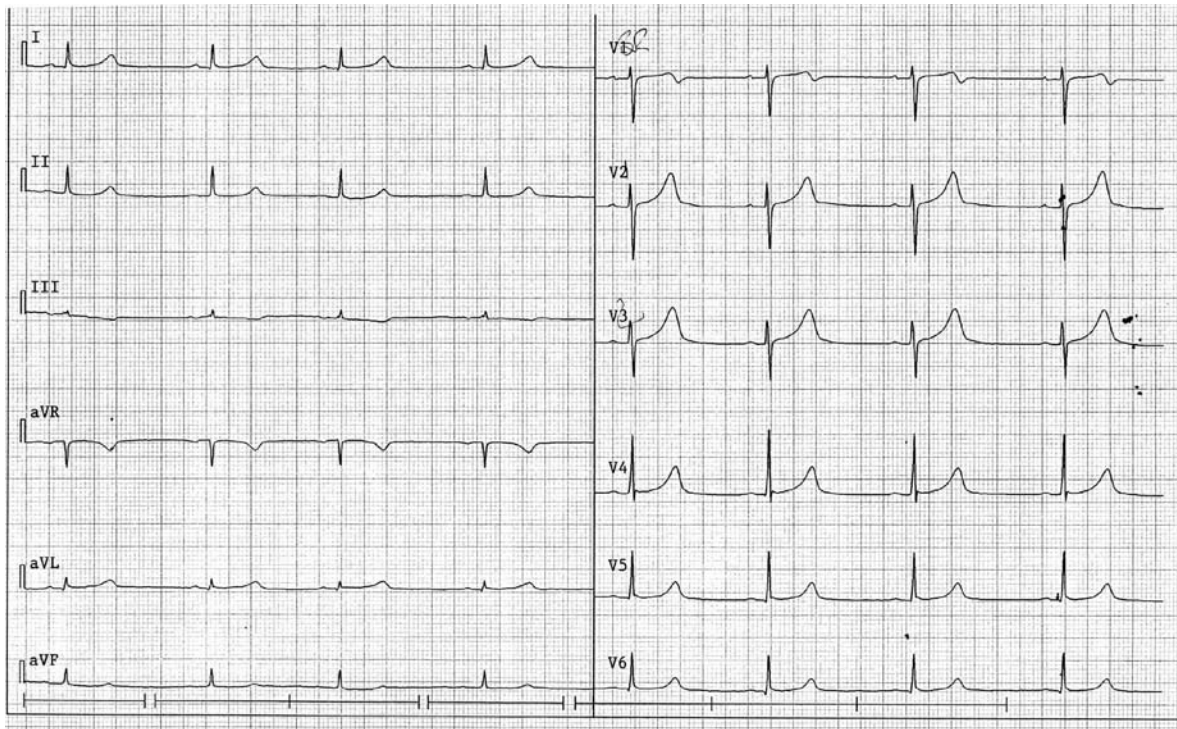
8. What is the next most appropriate action? (3 marks)

SELECT ONE ANSWER ONLY

- A increase peak inspiratory pressure on ventilator
- B manipulate existing chest drain
- C needle thoracentesis right chest
- D replace chest drain
- E reposition endotracheal tube

Question 9

A 15 year old boy presented with short self-limiting episodes of syncope. They occurred when he went to clubs with his friends.



25 mm / sec 10 mm / Mv

9. What significant abnormality is shown on his resting ECG? (3 marks)

SELECT ONE ANSWER ONLY

- A complete heart block
- B first degree heart block
- C left bundle branch block
- D left ventricular hypertrophy
- E prolonged Q-T interval
- F right bundle branch block
- G sinus arrhythmia
- H sinus bradycardia
- I sinus rhythm
- J Wolff-Parkinson-White syndrome

Questions 10, 11 and 12

This is a list of diagnoses:

- A aortic stenosis
- B breath holding attack
- C complex partial seizures ("petit mal")
- D fabricated seizure
- E febrile seizure
- F hypoglycaemia
- G juvenile myoclonic epilepsy
- H long QT syndrome
- I reflex anoxic seizures
- J rolandic seizures

Choose the most likely diagnosis for each of the following scenarios:

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

10. The mother of an 8 year old girl with type I diabetes gives a history of her daughter having shaking episodes during her sleep between 6 and 7am. During the episodes the girl is unresponsive and afterwards takes at least 30 minutes to get back to her normal self. Her blood sugar was 3.5 mmol/l during the most recent episode. Her parents have recently separated; she has a cousin on treatment for epilepsy. (3 marks)
11. A 2 year old girl is referred to outpatients for investigation of 2 episodes of unconsciousness. She becomes pale and shakes her arms and legs. One of these episodes happened when she fell off her tricycle on a day when she also had a mild elevation of temperature. The second episode occurred when another child pinched her at nursery. She has no significant previous medical history and her development is normal. (3 marks)
12. A 14 year old boy with type 1 diabetes presents with a series of collapses at school. His teacher provides a written history of the boy "going down like a sack of potatoes". The boy has no recall of events during these collapses. His neurological, cardiac examination and blood pressure are all normal. (3 marks)

Questions 13

A 6 week old breast fed baby girl presented with vomiting, poor feeding and lethargy of 1 day duration. On examination she was noted to have a capillary refill time of 4 seconds, temperature of 39°C and to be jaundiced, and her spleen was just palpable. Blood cultures have grown E. coli and she responded well to intravenous antibiotics, but the jaundice persisted.

She was born at term weighing 2.95 kg and was investigated for neonatal convulsions associated with hypoglycaemia which settled after the 1st week. She had been discharged home aged 10 days. Current investigation showed a total bilirubin of 159 µmol/l (conjugated 56 µmol/l).

13. Which one of the following additional investigations is most likely to help establish the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A brain MRI
- B galactose-1-phosphate uridyl transferase level
- C glucuronyl transferase level
- D hepatitis serology
- E HIDA liver scan
- F sweat test
- G thyroid function tests
- H ultrasound of abdomen
- I urinary steroid profile

Question 14

A 10 day old girl is admitted with 15% weight loss. She was born at term weighing 3.4 kg and has been breast fed since birth. On examination she appears well apart from obvious signs of dehydration.

Investigations

Blood

Sodium	160 mmol/l
Potassium	4.0 mmol/l
Urea	9.3 mmol/l
Creatinine	78 mmol/l
Glucose	4.4 mmol/l

She is started on normal saline at 15 mls/kg/hr. Four hours later her sodium is 161 mmol/l.

14. Which of the following is the appropriate fluid management at this stage? (4 marks)

SELECT TWO ANSWERS ONLY

- A change to 0.45% saline
- B change to 3% saline
- C change to 5% dextrose
- D decrease the rate of IV fluids
- E increase the rate of IV fluids
- F make no changes and repeat electrolytes in 4 hours

Questions 15 and 16

A 12 week old baby was found by his parents blue and apparently lifeless in his cot after they had heard him choking. His father revived him with mouth-to-mouth resuscitation and brought him to hospital.

He was born at 36 weeks gestation weighing 2.4 kg. He had no perinatal problems and fed well by bottle although he frequently regurgitated his feeds. His parents reported that he had been unwell with a cough for a few days before admission. His two siblings aged 5 years and 3 years had both been unwell recently with chest infections. The father had been unemployed for 18 months and there were considerable financial problems.

On examination he was pale and had a nasal discharge. His weight was on the 10th centile. Slight intercostal recession and a respiratory rate of 40/minute were noted together with scattered fine crepitations and a high pitched expiratory wheeze. The remainder of the examination was normal. Examination of the urine revealed no protein or sugar and no cells were seen on microscopy. A chest x-ray showed a degree of overinflation with no localised opacities.

15. What is the most useful investigation for establishing the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A echocardiogram
- B nasopharyngeal aspirate for respiratory viruses
- C pernasal swab
- D pH study
- E skeletal survey

This question continues on the next page.

16. What is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A congenital cardiac abnormality
- B cystic fibrosis
- C milk aspiration pneumonitis
- D pertussis infection
- E viral bronchiolitis

Question 17

An ex-preterm infant of 26 weeks gestation who is now 54 days old develops abdominal distension associated with temperature instability. An x-ray of the abdomen was performed which is shown below.



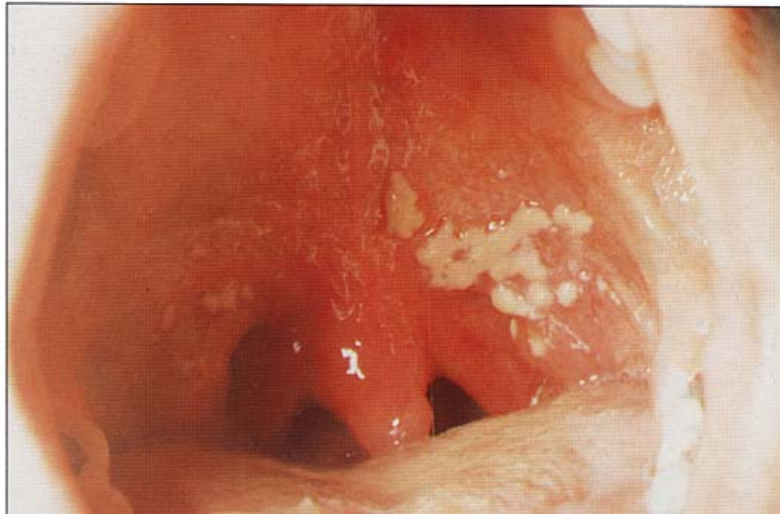
17. What is the next action in his management? (4 marks)

SELECT ONE ANSWER ONLY

- A Arrange for an upper gastrointestinal contrast study
- B Insert a nasogastric tube on free drainage
- C Intubation and ventilation
- D Rectal biopsy
- E Septic screen
- F Stop feeds and start IV antibiotics
- G Ultrasound scan of abdomen
- H Urgent referral to paediatric surgical team

Questions 18 and 19

This 16 year old girl presented with a sore throat.



18. What is the lesion on the soft palate? (3 marks)

SELECT ONE ANSWER ONLY

- A drug reaction
- B Epstein-Barr virus infection
- C Herpes simplex infection
- D oral Candidiasis
- E Staphylococcal infection
- F Streptococcal infection

This question continues on the next page.

19. Give three possible predisposing factors. (3 marks)

SELECT THREE ANSWERS ONLY

- A anti-fungal therapy
- B chemotherapy
- C Chlamydia infection
- D cow's milk allergy
- E Cytomegalovirus infection
- F gastro-oesophageal reflux
- G HIV infection
- H inhaled corticosteroid therapy
- I vitamin D deficiency
- J zinc deficiency

Questions 20, 21 and 22

This is a list of diagnoses:

- A achondroplasia
- B choanal atresia
- C Conradi syndrome (Rhizomelic chondrodysplasia punctata)
- D diastrophic dysplasia
- E hypochondroplasia
- F Jeune's asphyxiating thoracic dystrophy
- G myotonic dystrophy
- H osteogenesis imperfecta
- I Prader-Willi syndrome
- J pseudo-achondroplasia

Choose the most likely diagnosis for each of the following clinical scenarios:

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

20. A female infant is born at term weighing 3.2 kg. At birth she is noted to have a narrow chest and short limbs. She cries immediately, but develops rapid respiratory distress with virtually absent thoracic movements and becomes ventilator dependent. (3 marks)
21. A female infant is born at term weighing 2.9 kg. At birth she is noted to have a large head and very short bowed limbs. She cries immediately after birth, but then develops rapid respiratory distress with virtually absent thoracic movements and becomes ventilator dependent. (3 marks) *same pic of achondro but with bowed legs*
22. A female infant is born at term weighing 3.2 kg. At birth she is noted to have a large head, mid-face hypoplasia and relatively short limbs. She cries immediately after birth, but then develops mild respiratory distress which resolves within 24 hours. (3 marks)

Question 23

Study Incidence of bacteremia in infants and children with fever and petechiae.

Methods Consecutive patients with a temperature of 38° C or higher and petechiae seen in the emergency department were prospectively enrolled. Our measures included (1) laboratory tests (leukocyte count, coagulation profile, blood culture, and cerebrospinal fluid bacterial culture); (2) a questionnaire requesting clinical data including general appearance, number and location of petechiae, and presence or absence of purpura; and (3) a follow-up telephone survey documenting health status. **Results** A total of 411 patients were enrolled, with 57.7% between 3 and 36 months of age. Eight patients (1.9%) had bacteremia or clinical sepsis. None of the 357 well-appearing patients (95% confidence interval: 0.0% - 1.0%) had serious invasive bacteremia. 53 patients appeared ill, including all 6 with serious invasive bacteremia. Ill appearance of the child had a sensitivity of 1.00 (95% confidence interval: 0.60 - 1.00) and a leukocyte count of 15,000 or greater, or of less than 5000, had a sensitivity of 1.0 (95% confidence interval: 0.53 - 1.00) for detecting serious invasive bacteremia. All children with bacteremia had purpura.

Adapted from: Mandl KD, Stack AM, Fleisher GR. J Pediatr 1997 Sep;131(3):398-404

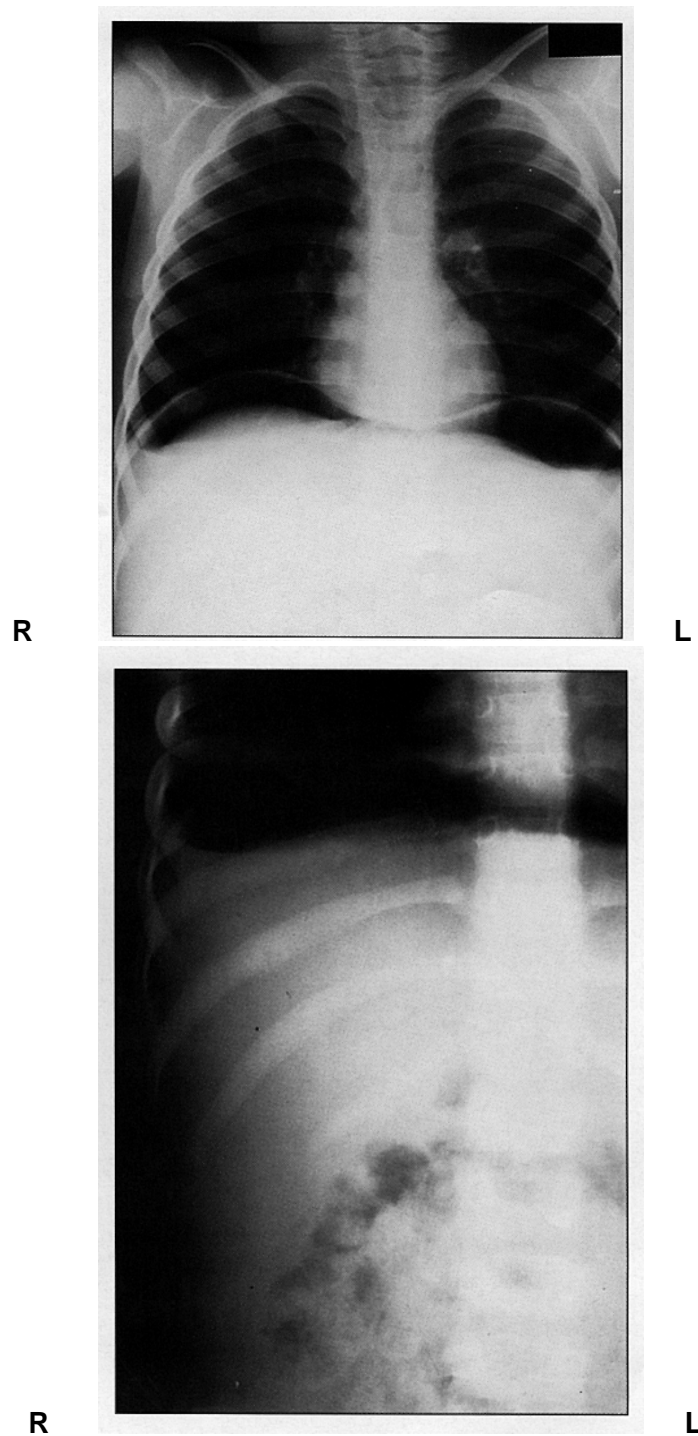
23. Which of the following statements is the most reasonable conclusion to be drawn from this study? (4 marks)

SELECT ONE ANSWER ONLY

- A All febrile children with petechiae should have a white blood cell count performed.
- B Assessment of whether the child looks ill is a highly sensitive test for serious invasive bacteraemia.
- C Assessment of whether the child looks ill is a highly specific test.
- D Children with a white cell count between 5,000 and 15,000 should be allowed home.
- E The white cell count is an accurate test for bacteraemia.
- F These results do not apply to the UK because the study was performed in the United States.

Question 24

These x-rays were taken of an 18 month old child on admission.



This question continues on the next page.

24. What abnormality is shown? (2 marks)

SELECT ONE ANSWER ONLY

- A ascites
- B bilateral sub-diaphragmatic air
- C fractured clavicle
- D hyperinflation
- E pneumomediastinum
- F pneumothorax
- G spina bifida occulta
- H subphrenic abscess

Question 25

A child with known congenital heart disease presents with this lesion.



25. What is the underlying diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A deep venous thrombosis
- B Factor V Leiden deficiency
- C idiopathic thrombocytopenic purpura
- D Meningococcal septicaemia
- E protein S deficiency
- F subacute bacterial endocarditis
- G systemic lupus erythematosus

Questions 26, 27 and 28

The following is a list of anti-epileptic drugs:

- A carbamazepine
- B diazepam
- C lamotrigine
- D lorazepam
- E paraldehyde
- F phenobarbitone
- G phenytoin
- H sodium valproate
- I topiramate
- J vigabatrin

Which drug would you use as a first line treatment in the following cases?

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

26. A term newborn with birth asphyxia presents at 12 hours of age with frequent short-lived generalised convulsions. (3 marks)
27. A 16 year old girl doing her GCSE exams with generalised tonic epilepsy. (3 marks)
28. A 7 year old boy with generalised absence seizures. (3 marks)

Questions 29 and 30

An 8 month old male infant is referred because of non-bilious vomiting. His GP had seen him frequently for constipation over the last few months.

Examination reveals a thin, non-dysmorphic infant weighing 6.8 kg (0.4th centile). He has a scaphoid abdomen and his capillary refill time is 3 seconds. General examination was otherwise unremarkable.

Investigations

Blood

haemoglobin	12.2 g/dl
white cell count	$13 \times 10^9/l$
neutrophils	$9.4 \times 10^9/l$
lymphocytes	$3.6 \times 10^9/l$
platelets	$373 \times 10^9/l$
sodium	154 mmol/l
potassium	3.8 mmol/l
urea	6.0 mmol/l

Urine

microscopy	no red cells no white cells no casts
osmolality	180 mOsm/kg

29. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A chronic renal failure
- B diabetes insipidus
- C gastro-oesophageal reflux
- D Hirschprung disease
- E renal tubular acidosis
- F type 1 diabetes
- G urinary tract infection

Question 30

30. Which of the following is the most appropriate test to confirm the diagnosis?
(3 marks)

SELECT ONE ANSWER ONLY

- A blood sugar
- B oesophageal pH study
- C plasma aldosterone level
- D plasma osmolality
- E rectal biopsy
- F response to DDAVP
- G urinary pH

Questions 31 and 32

A 14 year old boy of Bangladeshi origin was seen in the emergency department with a generalised convulsion. His parents said that he had complained for 2 weeks of mild headaches which had occurred at different times of the day. At the age of 12 he was found to be sniffing glue, but subsequently told his parents he had discontinued the practice. His progress at school was good and his behaviour had been normal.

On the afternoon of admission he had complained of a sudden generalised headache, despite this he had gone to see some friends but returned home with the headache. His mother had given him paracetamol. As he was sitting down to watch television, he became stiff and had a generalised convulsion.

The family called an ambulance and rectal diazepam was administered. He continued to fit and on arrival at hospital, intravenous lorazepam was required to terminate the convulsion. He remained very drowsy and non-responsive.

[important point](#)

On examination there was some resistance to flexion of his neck, but he was afebrile. His respirations were laboured; he was not cyanosed and was well perfused peripherally. His blood pressure was 160/90 mmHg. Examination of his heart, respiratory system and abdomen were normal.

His pupils were of equal size and both reacted sluggishly to light. Examination of the fundi showed no abnormalities. There was generalised increase in tone in his limbs, but no focal neurological signs.

31. What is the most appropriate investigation to establish the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A CT head scan
- B lumbar puncture
- C Mantoux test
- D toxicology screen
- E urgent EEG

This question continues on the next page.

32. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A cerebral tumour
- B drug overdose
- C epilepsy
- D meningitis
- E subarachnoid haemorrhage

Question 33

A 10 year old Somali girl developed an abscess in the wound of a compound fracture from which *E. coli* was cultured.

Investigations

Blood

haemoglobin	6.8 g/dl
MCV	82.3 fl
PCV	0.22
MCHC	30.6 g/dl
white cell count	$12.3 \times 10^9/l$
neutrophils	$9.1 \times 10^9/l$
lymphocytes	$2.3 \times 10^9/l$
monocytes	$0.9 \times 10^9/l$
reticulocytes	5.5%
platelets	$79 \times 10^9/l$
blood film	anisocytosis with microcytes and burr cells
Coombs test	negative

Urine microscopy	$200 \text{ red blood cells } \times 10^6/l$ granular casts
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33. What is the most likely cause of the anaemia? (4 marks)

SELECT ONE ANSWER ONLY

- A acute lymphoblastic leukaemia
- B blood loss
- C HIV infection
- D malaria
- E microangiopathic intravascular haemolysis
- F sickle cell disease
- G thalassaemia
- H tuberculosis

Question 34

A 13 year old girl with a 3 year history of type I diabetes presents to the clinic with poor control and a history of weight loss for 3 months.

During this time her control has gradually deteriorated without obvious reason. Her total insulin dose has progressively increased to over 60 units daily and she has been changed on to a 4 injection per day regimen. Her record book shows few blood results but most are within the normal range. She complains of thirst and nocturia, mild intermittent abdominal pain and her weight has fallen from 45 kg to 40 kg.

She is the second of three daughters of a stable family with no financial worries. She had been successful and happy at school, but recent events have caused school absences.

On examination she is tall, height 162cm (75-90th centile) and thin, weight 40 kg (<25th centile). General examination is otherwise unremarkable. Her injection areas are not hypertrophied. She is at puberty stage 2.

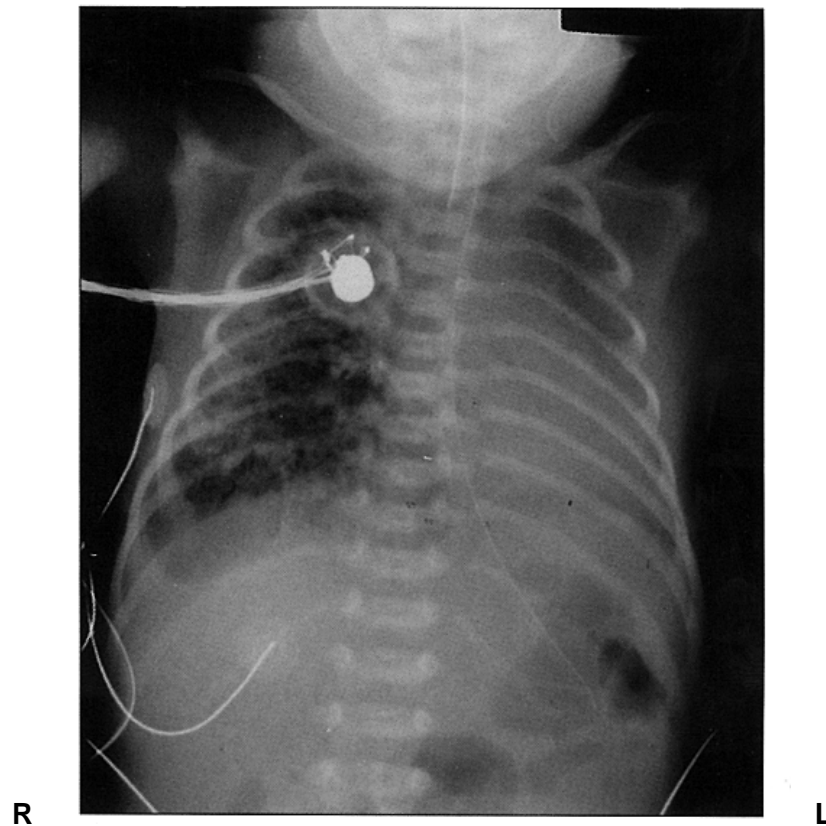
34. What is the most likely cause of her recent weight loss? (2 marks)

SELECT ONE ANSWER ONLY

- A anorexia nervosa
- B coeliac disease
- C hyperthyroidism
- D occult urinary tract infection
- E poor compliance with diet
- F poor compliance with insulin

Question 35

This is the x-ray of a 7 day old baby with severe respiratory difficulty.



35. What is the most likely diagnosis of the abnormal right lung? (3 marks)

SELECT ONE ANSWER ONLY

- A bronchogenic cyst
- B congenital lobar emphysema
- C cystic adenomatoid malformation
- D diaphragmatic hernia
- E MacLeod's syndrome
- F meconium pneumonitis
- G pneumothorax
- H pulmonary interstitial emphysema

I Questions 36 and 37

The dentist reported this incidental finding in a 16 year old female.



36. What is the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A aberrant parathyroid
- B cavernous haemangioma
- C cystic hygroma
- D lingual thyroid
- E lymphoma
- F mucus retention cyst
- G peri-tonsillar abscess

This question continues on the next page.

37. How would you confirm the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A chest x-ray
- B excision biopsy
- C MRI scan of head and neck
- D needle aspiration
- E parathyroid hormone levels
- F thyroid scan

Questions 38, 39 and 40

This is a list of clinical diagnoses:

- A Addison's disease
- B aspirin toxicity
- C cerebral oedema
- D diabetic ketoacidosis
- E febrile convulsion
- F glycogen storage disease
- G hypopituitarism
- H medium chain acyl-coA dehydrogenase deficiency (MCAD)
- I pyridoxine dependent seizures
- J Reye's syndrome

Match the following case descriptions with the most likely diagnosis from the list above:

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each option may be used more than once*

38. A 13 year old girl has had poor diabetic control and has been feeling less well. She is admitted with diabetes ketoacidosis. Initial blood glucose is 27 mmol/l with a pH of 7.0. She responds well to fluids, resuscitation and insulin and her blood glucose falls to 10 mmol/l. Twelve hours after admission her level of consciousness deteriorates and her pulse drops to 70/min. (3 marks)
39. A 2 year old boy becomes coryzal. He vomits three times and subsequently becomes drowsy, followed by a generalised tonic clonic seizure. Blood glucose 4.6 mmol/l, sodium 137 mmol/l, potassium 4.5 mmol/l, creatinine 30 mmol/l, urea 2.5mmol/l. (3 marks)
40. A 4 month old male infant is admitted with a generalised seizure lasting 10 minutes. The seizure is eventually terminated with rectal diazepam. On examination he is noted to have a liver palpable 5 cm below the costal margin. Blood glucose 1.8 mmol/l. (3 marks)

Question 41

A 10 year old girl presented because of faltering growth and 4 months of ill-defined abdominal pain. On examination her height was 128 cm (9th centile) and weight 22 kg (2nd centile). She had several mouth ulcers and appeared pale, but examination was otherwise normal.

Investigations

Blood

haemoglobin	9.6 g/dl
MCH	26 pg
MCV	76 fl
white cell count	$6.6 \times 10^9/l$
platelets	$500 \times 10^9/l$
CRP	22 mg/l
ferritin	15 $\mu g/l$ (15-250)

41. What is the most likely cause of her problems? (5 marks)

SELECT ONE ANSWER ONLY

- A coeliac disease
- B Crohn's disease
- C eosinophilic colitis
- D iron deficiency anaemia
- E psychogenic pain
- F ulcerative colitis

Question 42

A 5 month old boy has had several hospital admissions with episodes of reduced conscious level, altered muscle tone and abnormal movements. These last for several hours and have remained unexplained despite extensive in-patient investigation. He is found cyanosed and apnoeic whilst being cared for by his mother in a cubicle. He responds rapidly to basic life support. Continuous nursing supervision is arranged.

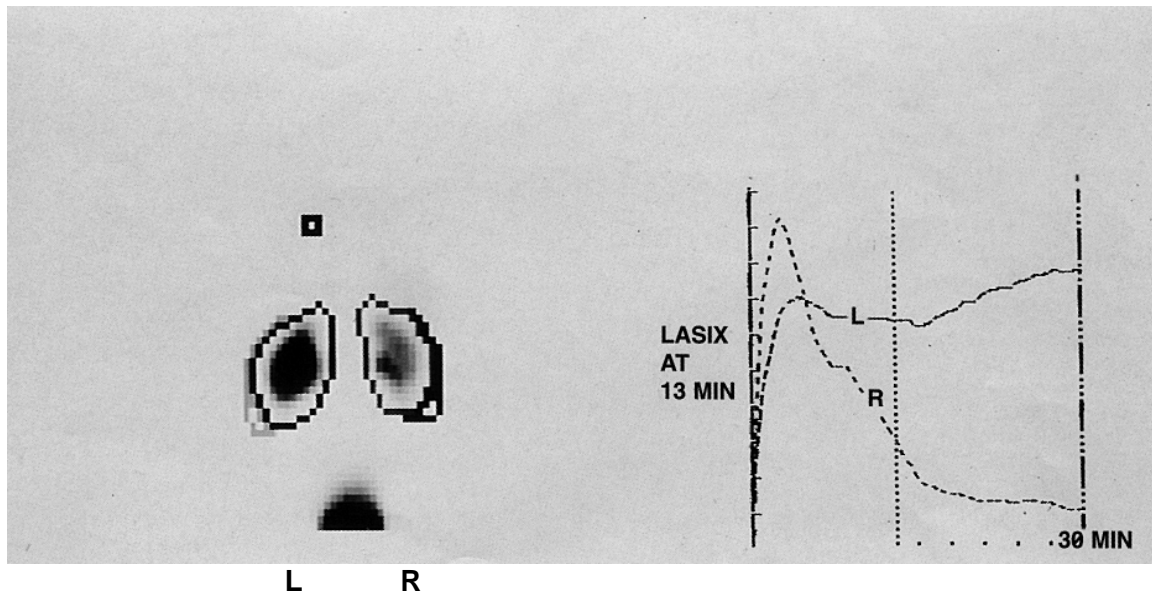
42. What is the most appropriate next step? (4 marks)

SELECT ONE ANSWER ONLY

- A Arrange a skeletal survey.
- B Confront the mother about your suspicions.
- C Refer to social services.
- D Repeat full infection screen.
- E Seek an immediate Police Protection Order.

Questions 43 and 44

This is a MAG 3 renogram on a boy with recurrent urinary tract infection.



43. What important structural abnormality is shown? (2 marks)

SELECT ONE ANSWER ONLY

- A bilateral pelvi-ureteric junction obstruction
- B bilateral renal cortical scarring
- C dilated left renal pelvis
- D dilated right renal pelvis
- E left renal cortical scarring
- F right pelvi-ureteric obstruction
- G right renal cortical scarring
- H posterior urethral valves

This question continues on the next page.

44. What functional abnormality is shown? (2 marks)

SELECT ONE ANSWER ONLY

- A bilaterally delayed excretion
- B bilateral ureteric reflux
- C delayed excretion on left despite furosemide
- D delayed excretion on right despite furosemide
- E left ureteric reflux
- F poor uptake on the left
- G poor uptake on the right
- H right ureteric reflux

Question 45

This 3 year old Romanian child, who has recently arrived in the UK, was noted to have an abnormality of his nail bed.



45. In which of the following is this physical sign a clinical feature? (3 marks)

SELECT ONE ANSWER ONLY

- A congenital cystic adenomatoid malformation
- B Eisenmenger syndrome
- C homozygous sickle cell disease
- D polycystic kidneys
- E pulmonary stenosis

Question 46

A 15 year old girl with a BMI of 15 has been diagnosed with ileocaecal Crohn's disease. She is to be commenced on enteral nutrition given by nasogastric tube in order to achieve remission.

Investigations

Blood

haemoglobin	9.3 g/dl
white cell count	11.3 x 10 ⁹ /l
normal differential	
platelets	275 x 10 ⁹ /l
ferritin	15 µg/l (15-300)
sodium	133 mmol/l
chloride	95 mmol/l
potassium	2.9 mmol/l
urea	6.0 mmol/l
creatinine	55 µmol/l
phosphate	1.3 mmol/l

46. Which of the following is the most important action to take before commencing the enteral feeding regime? (4 marks)

SELECT ONE ANSWER ONLY

- A commence oral iron therapy
- B correct hypokalaemia
- C give intramuscular vitamin B12
- D start oral multivitamin therapy
- E transfuse haemoglobin to 12 g/dl

Question 47

A 2 year old Asian boy was noted to be pale by his GP while being examined for a respiratory tract infection. Full blood count showed haemoglobin of 3.2 g/dl. He was born at term with no perinatal problems and was still being breast fed.

47. What is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A coeliac disease
- B hereditary spherocytosis
- C iron deficiency anaemia
- D Meckel's diverticulum
- E thalassemia major

Question 48

This is the x-ray of an 18 month old child referred because of decreased movement of the left arm.



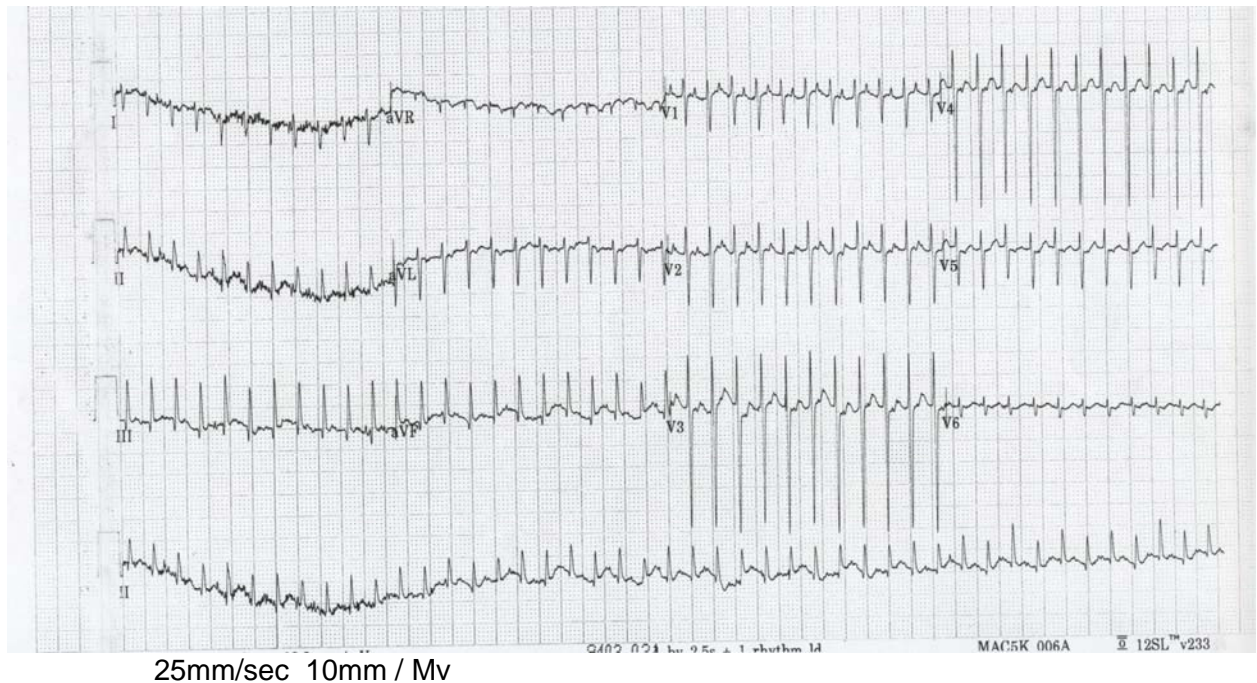
48. What abnormalities are shown in the x-ray? (4 marks)

SELECT TWO ANSWERS ONLY

- A fractures of radius and ulna
- B lead poisoning
- C mucopolysaccharidosis
- D non-accidental injury
- E osteogenesis imperfecta
- F osteomyelitis
- G osteopetrosis
- H pseudohypoparathyroidism
- I rickets

Question 49

A 2 week old baby boy, born at term and previously well, presented to the emergency department with drowsiness, poor feeding and rapid breathing. On examination he was pale, shocked, breathless and generally unwell. Oxygen saturation was 96% in air and he had a heart rate of 270/minute. His ECG is shown below.



49. Which of the following is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A atrial flutter
- B atrial tachycardia with 2:1 block
- C supraventricular tachycardia → narrow QRS complex and no P wave
- D ventricular fibrillation
- E ventricular tachycardia

Question 50

A 12 year old boy is noted to be short. His mother is concerned about his short stature but he is not. On examination he has Tanner axillary hair stage 1, pubic hair stage 1, genitalia stage 1 and testicular volumes 2 ml bilaterally. His bone age is 9.5 years.

50. Which is the most appropriate management option? (4 marks)

SELECT ONE ANSWER ONLY

- A high dose testosterone for 4 months
- B low dose testosterone treatment for 4 months
- C reassure and discharge
- D reassure and follow up in 4 months
- E recombinant Growth Hormone treatment

Question 51



51. Select the three most likely causes for the appearance of this 4 year old boy. (3 marks)

SELECT THREE ANSWERS ONLY

- A acute nephritis
- B bilateral ethmoid sinusitis
- C bilateral periorbital cellulitis
- D cardiac failure
- E cavernous sinus thrombosis
- F congenital angio-oedema
- G Henoch-Schönlein purpura
- H myotonia
- I nephrotic syndrome
- J non-accidental injury
- K superior vena cava obstruction
- L water intoxication

Question 52

An 18 month old Caucasian girl presented with a 3 week history of screaming at night. Over the past week she had developed a swelling in her neck. Her 3½ year old brother had a history of Toxoplasmosis in the past. There was no family history of tuberculosis. The child was well with a good appetite, no fever and clinical examination revealed a lump at the angle of her jaw. Ear, nose and throat examination was normal. There was no other lymphadenopathy and no hepatosplenomegaly.

52. Which of the following treatments would be the most appropriate? (4 marks)

SELECT ONE ANSWER ONLY

- A intravenous cefotaxime
- B oral clarithromycin
- C oral flucloxacillin and amoxicillin
- D oral prednisolone
- E oral rifampicin and isoniazid

Question 53

A 2 year old boy was referred with a history of delay in walking and poor speech development. He was described as “if in a world of his own”. He had been born normally at term with a birth weight of 3.8 kg. He was very sensitive to noise. His development was described as normal up to the age of 9 months. Following a viral upper respiratory tract infection his parents noted that he was not as communicative as before.

On examination he had a slightly upturned nose, low nasal bridge and a long philtrum. Wood's lamp examination of his skin was normal. Muscle tone was generally reduced but reflexes were all present. Both heart sounds were normal, but there was a grade 3/6 systolic murmur at the upper left sternal edge.


Investigations

Blood

full blood count	normal
sodium	137 mmol/l
potassium	3.9 mmol/l
calcium	2.92 mmol/l
urea	5.0 mmol/l
CPK	70 U/l (60-300)

53. Which of the following is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A autistic spectrum disorder
- B Di-George syndrome
- C Duchenne muscular dystrophy
- D fetal alcohol syndrome
- E fragile X syndrome
- F Williams syndrome  associated with autistic

Question 54

An 8 year old girl was diagnosed as having cystic fibrosis on neonatal screening. She had been treated with oral flucloxacillin for intermittent *Staphylococcus aureus* grown from cough swabs in the clinic. The most recent sample has grown *Pseudomonas aeruginosa*.

54. Which of the following is the most appropriate management? (3 marks)

SELECT ONE ANSWER ONLY

- A inhaled tobramycin for 3 weeks
- B IV ceftazidime for 14 days
- C nebulised Dnase for 3 weeks
- D nebulised hypertonic saline for 3 weeks
- E oral ciprofloxacin and nebulised colomycin for 3 months

Question 55

An 8 year old boy presented with acute lymphoblastic leukaemia a year ago. He has relapsed following a bone marrow transplant and there are no other curative options. He has told the palliative care nurse that "I don't want any more hospital". Over the last week he has become weak and drowsy, not being able to leave his bed. His parents asked for a blood test which shows haemoglobin of 7.8 g/dl. His parents request a blood transfusion. It is the view of the paediatric oncology team that he should not be transfused.

55. What is the rationale of the oncology team for this opinion? (4 marks)

SELECT ONE ANSWER ONLY

- A He is Fraser (Gillick) competent.
- B His parents are too distressed to understand the prognosis.
- C It is contraindicated in the shared-care guidelines.
- D Treatment simply delays death.
- E Treatment will not improve the symptoms.

Question 56

This is an x-ray of the abdomen of a 12 year old girl who attends a school for children with learning difficulties and who complains of recurrent abdominal pain.



56. What abnormality can be seen on the plain abdominal film? (3 marks)

SELECT ONE ANSWER ONLY

- A bilateral renal calculi
- B bilateral sacro-ileitis
- C calcification of the pancreas
- D calcified para-aortic lymph nodes
- E constipation
- F megacolon
- G nephrocalcinosis

Question 57

A 4 year old child presented with an abdominal mass.



57. What two abnormalities are seen on the CT scan with IV contrast? (4 marks)

SELECT TWO ANSWERS ONLY

- A displaced right kidney
- B inferior vena cava thrombosis
- C megacolon
- D multi-cystic right kidney
- E pancreatic pseudocyst
- F para-aortic lymph node enlargement
- G solid tumour of the liver
- H solid tumour of the right adrenal gland
- I solid tumour of the right kidney
- J spina bifida occulta

Questions 58, 59 and 60

The following is a list of investigations for infants with scrotal problems:

- A 17-hydroxyprogesterone assay
- B 21-alpha-hydroxylase assay
- C CT abdomen
- D DNA analysis for CYP21 gene mutation
- E GP review at 6 months
- F karyotyping
- G referral to clinical geneticist
- H referral to paediatric surgeon
- I ultrasound of abdomen
- J urea and electrolytes

Which would be the next step in the immediate management of each of the following patients?

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once.*

- 58. At 6 week check this baby was noted to have an empty scrotum. A mobile non-tender mass is palpable high in the inguinal canal. (3 marks)
- 59. At 6 week check this baby was noted to have an empty scrotum and ^{U&E}hypospadias. (3 marks)
- 60. At 6 week check this baby was noted to have a firm tender swelling in the right inguinal canal. Both testes are in the scrotum. (3 marks)

End of Specimen Paper

Royal College of Paediatrics and Child Health

APPLIED KNOWLEDGE IN PRACTICE

PAPER 2

Q No	No of Ans	Ans	Marks	Answer in words
1	1	E	3	offer reassurance
2	1	H	3	pertussis
3	1	F	3	habitual cough
4	1	I	3	primary ciliary dyskinesia
5	1	B	4	low maternal platelet count in pregnancy
6	1	D	3	reassure child and review him in 6 months time
7	2	D	2	Short course treatment resulted in a significantly reduced length of hospital stay
		F	2	There was no difference in the level of neurological complications between treatment groups.
8	1	E	3	reposition endotracheal tube
9	1	E	3	prolonged Q - T interval
10	1	J	3	rolandic seizures
11	1	I	3	reflexic anoxic seizure
12	1	H	3	long Q-T syndrome
13	1	B	3	galactose-1-phosphate uridyl transferase level
14	2	A	3	change to 0.45% saline
		E	3	increase the rate of IV fluids
15	1	B	3	nasopharyngeal aspirate for respiratory viruses
16	1	E	4	viral bronchiolitis
17	1	F	4	Stop feeds and start IV antibiotics
18	1	D	3	oral Candidiasis
19	3	B	1	chemotherapy
		G	1	HIV infection
		H	1	inhaled corticosteroid therapy
20	1	F	3	Jeune's asphyxiating thoracic dystrophy
21	1	H	3	osteogenesis imperfecta
22	1	A	3	Achondroplasia
23	1	B	4	Assessment of whether the child looks ill is a highly sensitive test for serious invasive bacteraemia.
24	1	B	2	bilateral sub-diaphragmatic air
25	1	F	3	subacute bacterial endocarditis
26	1	F	3	phenobarbitone
27	1	C	3	lamotrigine
28	1	H	3	sodium valproate
29	1	B	3	diabetes insipidus
30	1	F	3	response to DDAVP
31	1	A	3	CT head scan
32	1	E	3	subarachnoid haemorrhage
33	1	E	4	microangiopathic intravascular haemolysis
34	1	F	2	poor compliance with insulin
35	1	H	3	pulmonary interstitial emphysema
36	1	D	3	lingual thyroid
37	1	F	3	thyroid scan
38	1	C	3	cerebral oedema
39	1	E	3	febrile convulsion
40	1	F	3	glycogen storage disease
41	1	B	5	Crohn's disease
42	1	C	4	Refer to social services
43	1	C	2	dilated left renal pelvis
44	1	C	2	delayed excretion on left despite furosemide

Royal College of Paediatrics and Child Health

APPLIED KNOWLEDGE IN PRACTICE

PAPER 2

Q No	No of Ans	Ans	Marks	Answer in words
45	1	B	3	Eisenmenger syndrome
46	1	B	4	correct hypokalaemia
47	1	C	4	iron deficiency anaemia
48	2	A	2	fractures of radius and ulna
		I	2	rickets
49	1	C	3	supraventricular tachycardia
50	1	D	4	reassure and follow up in 4 months
51	3	A	1	acute nephritis
		F	1	congenital angio-oedema
		I	1	nephrotic syndrome
52	1	C	4	oral flucloxacillin and amoxicillin
53	1	F	3	Williams syndrome
54	1	E	3	oral ciprofloxacin and nebulised colomycin for 3 months
55	1	D	4	Treatment simply delays death
56	1	G	3	nephrocalcinosis
57	2	A	2	displaced right kidney
		I	2	solid tumour of the right kidney
58	1	E	3	GP review at 6 months
59	1	J	3	urea and electrolytes
60	1	H	3	Referral to paediatric surgeon

THE END

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This 6 month old baby boy has had a mild skin rash since the age of 2 months. He has suddenly become unwell with fever and marked worsening of the rash.

What is the most likely diagnosis? (4 marks)

Select one answer only

Clicking on the image below will open it up in a new window, allowing it to be moved around the screen

(C 1691 SP)



A Chickenpox (varicella)

B Eczema herpeticum

C Impetigo

D Primary Herpes Simplex infection

E Stevens-Johnson syndrome

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An 18 month old boy presents with a history of persistent cough for 4 weeks. He had a urinary tract infection 3 weeks previously, but had recovered with only symptomatic treatment.

On examination, his chest is clear to auscultation, his heart sounds are normal and his abdomen is soft with no organomegaly present. His chest x-ray is shown below.

What is the most likely diagnosis? (4 marks)

Select one answer only

Clicking on the x-ray below will open it up in a new window, allowing it to be moved around the screen

(X 0020 SP)



A Congenital diaphragmatic hernia

B Congenital lobar emphysema

C Congenital lung cyst

D Loculated pneumothorax

E Pneumatocele

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You will be presented with three clinical scenarios and a list of respiratory diagnoses.

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Type in one answer only for each of the 3 clinical scenarios below.

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Each question is worth 3 marks.

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Note: each answer may be used more than once.

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(EMQ 0007 SP)

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A 5 year old girl presents with a persistent cough following right lower lobe pneumonia when aged 4 years. Crackles are heard in the right base.

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Select the most likely diagnosis from the list below: (3 marks)

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A Bronchiectasis

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B Chlamydia pneumonitis

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C Gastro-oesophageal reflux

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D Hyper IgE syndrome

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E Obliterative bronchiolitis

F Pertussis

G Primary ciliary dyskinesia

H Pulmonary tuberculosis

I Recurrent aspiration

J Tracheo-oesophageal fistula

Please add the correct answer in the box then continue to scroll down to the next part of the question

(EMQ 0007 SP a)

A 2 year old boy who suffered from severe birth asphyxia presents with recurrent cough and wheeze. He had mild eczema as an infant.

Select the most likely diagnosis from the list below: (3 marks)

A Bronchiectasis

B Chlamydia pneumonitis



Back

Next

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C Gastro-oesophageal reflux

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D Hyper IgE syndrome

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E Obliterative bronchiolitis

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F Pertussis

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G Primary ciliary dyskinesia

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H Pulmonary tuberculosis

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I Recurrent aspiration

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J Tracheo-oesophageal fistula

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Please add the correct answer in the box then continue to scroll down to the next part of the question

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(EMQ 0007 SP b)

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A 3 year old boy presents with a history of recurrent skin sepsis, eczema, chronic cough and sputum production. Sputum samples grow *Staphylococcus aureus*.

Select the most likely diagnosis from the list below: (3 marks)

A Bronchiectasis

B Chlamydia pneumonitis

C Gastro-oesophageal reflux

D Hyper IgE syndrome

E Obliterative bronchiolitis

F Pertussis

G Primary ciliary dyskinesia

H Pulmonary tuberculosis

I Recurrent aspiration

J Tracheo-oesophageal fistula

Please add the correct answer in the box below

(EMQ 0007 SP c)

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A 14 year old girl presents with a 3 week history of facial rash and a similar rash over her shoulders following exposure to the sun. This was followed by increasing tiredness and malaise, swelling of the right thumb and the proximal interphalangeal joint of the right middle finger, shortness of breath on exertion and a dry cough. During the previous week, she had complained of left sided pleuritic pain. Her appearance is shown below.

On examination, her temperature is 37.8°C. She has a tender spot in the pulp of both thumbs. She has tachycardia, a pleuro-pericardial rub and crepitations at the left base. Test results are also shown below.

Which of the following investigations is most likely to lead to the diagnosis? (5 marks)

Select one answer only

Clicking on the image below will open it up in a new window, allowing it to be moved around the screen



Investigations:

Blood

haemoglobin	105 g/l	(115-165)
white cell count	$3.2 \times 10^9/l$	(3.0-10.0)
C-reactive protein	49 mg/l	(< 5 mg/l)
blood culture	awaited	

A Anti-double stranded DNA

B Mycoplasma titres

C Parvovirus titres

D Rheumatoid factor

E Skin swab

F Urinary porphyrin

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A 4 day old boy, born to consanguineous Asian parents, was brought to the emergency department with left sided focal fits. The fits stopped after giving rectal diazepam.

On examination, his temperature is 38°C, heart rate 140/minute and respiratory rate 40/minute. He is drowsy post diazepam but can be roused. The rest of the physical examination is normal.

Apart from a septic screen, including a lumbar puncture, which two of the following investigations would be most helpful in his immediate management? (8 marks)

Select two answers only

A Ammonia

B Blood sugar

C Calcium

D Coagulation screen

E Cranial ultrasound

F CT head scan

G Full blood count

H Urea and electrolytes

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A 2 year old boy presents with a history of recurrent episodes of colicky abdominal pain for 2 months. His abdomen appeared bloated at times. His bowel motions were regular, but he had passed a blood stained stool on 3 separate occasions. His ^{99m}Tc pertechnetate scan is shown below.

What is the most likely diagnosis? (5 marks)

Select one answer only

Clicking on the scan below will open it up in a new window, allowing it to be moved around the screen

(C 2064 SP)



A Appendix abscess

B Intussusception

C Malrotation

D Meckel's diverticulum

E Vesico-ureteric reflux

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This infant was referred from the midwifery led unit, having been born at term following an uneventful pregnancy. His clinical photograph is shown below.

What is the most important initial investigation? (4 marks)

Select one answer only

Clicking on the image below will open it up in a new window, allowing it to be moved around the page

(C 0005 SP)



A Chromosome analysis

B Cranial CT scan

C Cranial ultrasound

D EEG

E Intraocular pressure measurement

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A 4 month old baby boy from Eastern Europe who had recently arrived in the UK presents with poor feeding, loose stools and faltering growth.

On examination, his length and weight are on the 2nd centiles for age. He is noted to have oral candidiasis and a mild degree of bilateral parotid swelling. Test results are shown below.

What is the most likely diagnosis? (5 marks)

Select one answer only

Clicking on the data chart below will open it up in a new window, allowing it to be moved around the screen

(D 0003 SP)

Investigations:

Blood

haemoglobin	83 g/l	(110 - 140)
white cell count	$5.2 \times 10^9/l$	(6.0 - 15.0)
neutrophils	$3.1 \times 10^9/l$	(1.5 - 8.0)
lymphocytes	$2.0 \times 10^9/l$	(4.0 - 10.0)
platelets	$485 \times 10^9/l$	(150 - 450)

Immunoglobulins

IgG	9.5 g/l	(2.1 - 10.9)
IgA	1.2 g/l	(0.05 - 0.7)
IgM	1.8 g/l	(0.15 - 2.1)

A Chronic granulomatous disease

B Common variable immune deficiency

C HIV/AIDs

D Severe combined immune deficiency

E X-linked hypogammaglobulinaemia

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This 9 month old male infant presented with a 3 day history of fever, rhinorrhoea and rash. His mother had brought him to see the GP, who prescribed erythromycin and antipyretics. The child did not improve and his rash worsened, extending from his face to the rest of his body, in particular the axillae and groins.

What is the most likely diagnosis? (4 marks)

Select one answer only

Clicking on the image below will open it up in a new window, allowing it to be moved around the screen

(C 0007 SP)



A Atopic dermatitis

B Herpes simplex infection with secondary bacterial infection

C Impetigo

D Staphylococcal scalded skin syndrome

E Stevens-Johnson Syndrome

F Toxic epidermal necrolysis

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This 28 week gestation baby, currently ventilated, had a worsening metabolic acidosis over the last 24 hours.

On examination, his heart rate is 180/minute and mean blood pressure is 26 mmHg. He has developed peripheral oedema. Due to bilious aspirates an abdominal x-ray was performed.

What is the most appropriate immediate management? (5 marks)

Select one answer only

Clicking on the x-ray image below will open it up in a new window, allowing it to be moved around the screen

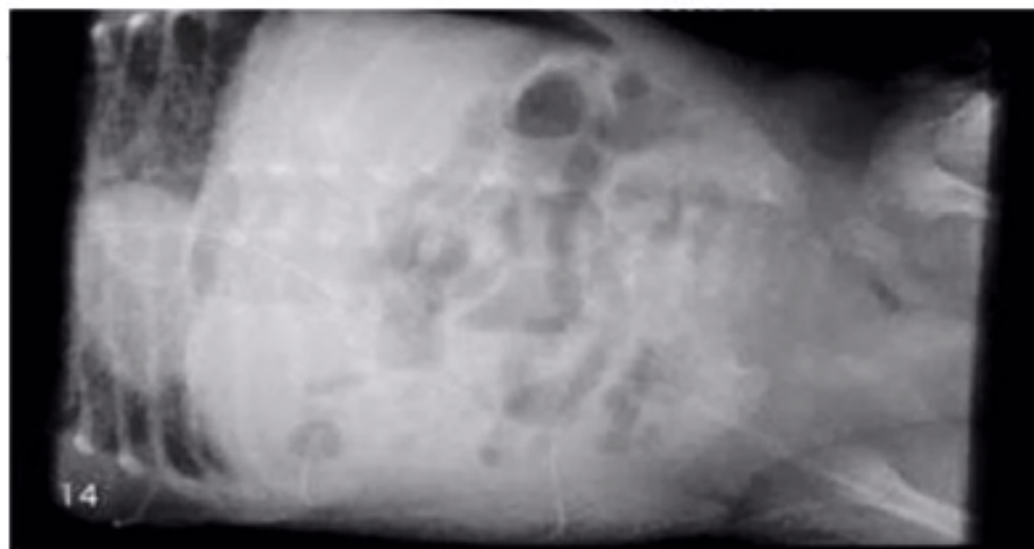
(X 0011 SP)

Investigations:

Blood gas

pH	7.01	(7.35 - 7.45)
pCO ₂	5.5 kPa	(4.6 - 6.4)
pO ₂	8.1 kPa	(11 - 15)
HCO ₃	10.2 mmol/l	(19 - 28)
BE	-17.0 mmol/l	(-2 to +2)

Here is his abdominal x-ray



A Administer 1 mmol/kg sodium bicarbonate

B Fluid resuscitation with 10 ml/kg 0.9% saline

C Increase ventilation pressures and rate

D Needle aspiration of abdomen

E Start intravenous antibiotics

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A 2 year old girl was referred to outpatients for investigation of 2 episodes of unconsciousness. Her mother described that the child became pale and both her arms and legs were seen to shake for about 45 seconds. She was unresponsive during the episode and was noted to be drowsy for a while afterwards.

One of the episodes occurred after she fell off her tricycle on a day when she also had a mild elevation of temperature. The second episode occurred when another child pinched her at nursery. She has no significant previous medical history and her development is normal. Examination in the outpatients department finds no abnormalities.

What is the most likely diagnosis? (5 marks)

Select one answer only

A Benign seizures of childhood

B Complex partial seizures

C Fabricated seizure

D Febrile seizure

E Hypoglycaemia

F Juvenile myoclonic epilepsy

G Reflex anoxic seizures



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This question consists of two parts. Please answer both parts of the question.

This is the growth chart of a 10 month old boy who was admitted to hospital because of vomiting and poor weight gain. The baby fed well on a mixed diet. The stools were normal. There were no abnormal findings apart from wasting.

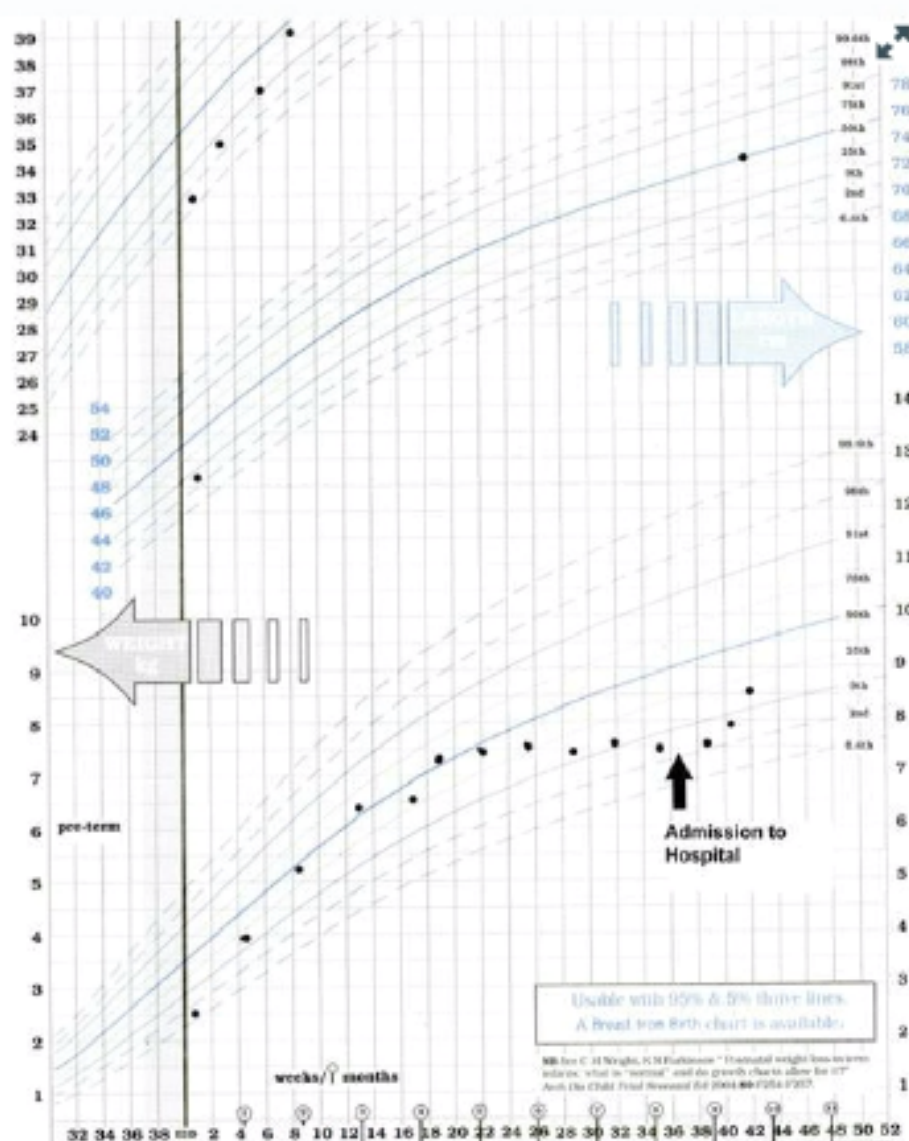
What is the most likely diagnosis? (5 marks)

Select one answer only

Clicking on the chart below will open it up in a new window, allowing it to be moved around the screen

Please continue to scroll down to the next part of the question

(C 0011 SP a)



A Coeliac disease

B Cows' milk protein intolerance

C Cystic fibrosis

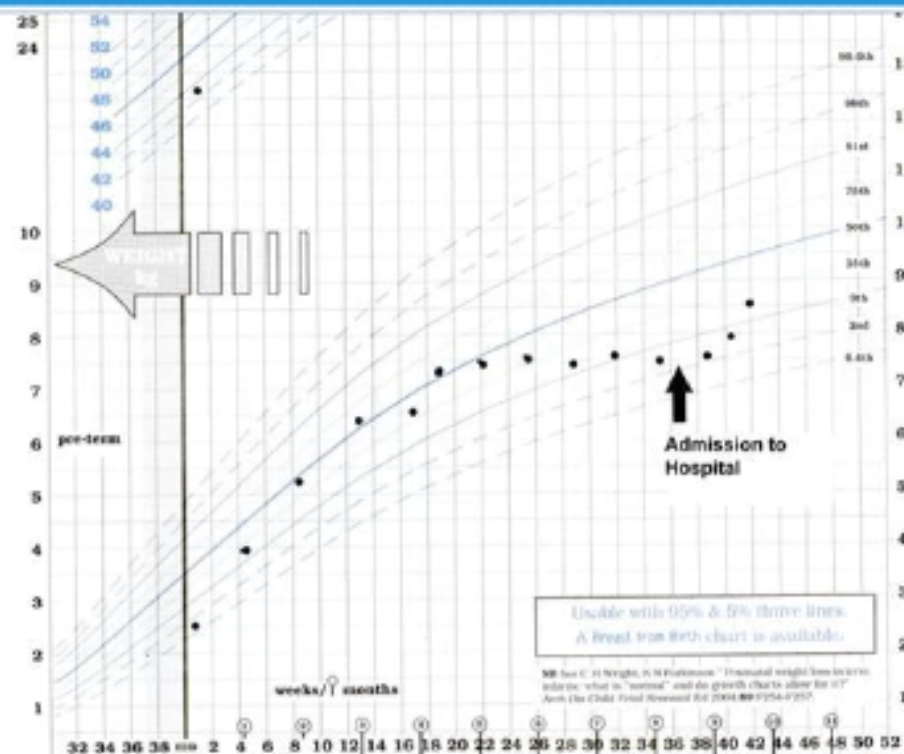
D Gastro-oesophageal reflux disease



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- A Coeliac disease
- B Cows' milk protein intolerance
- C Cystic fibrosis
- D Gastro-oesophageal reflux disease
- E Inadequate calorie intake

In light of this growth chart, what is the single most helpful next action? (4 marks)

Select one answer only

(C 0011 SP b)

- A Discharge planning meeting
- B Food diary for one week
- C Measure IgA anti-tissue transglutaminase antibodies
- D Oesophageal pH monitoring
- E RAST test for cows' milk IgE

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You will be presented with three clinical scenarios and a list of treatment options for seizures.

Type in one answer only for each of the 3 clinical scenarios below

Each question is worth 3 marks.

Note: each answer may be used more than once

(EMQ 0003 SP)

A 13 year old girl with juvenile absence epilepsy.

Choose the most appropriate treatment: (3 marks)

A Carbamazepine

B Lamotrigine

C Levetiracetam

D No treatment

E Phenobarbitone

F Phenytoin

G Sodium valproate

H Topiramate

I Vigabatrin

J Zonisamide

Please add the correct answer in the box then continue to scroll down to the next part of the question

(EMQ 0003 SP a)

A 6 month old boy with infantile spasms and developmental delay.

Choose the most appropriate treatment: (3 marks)

A Carbamazepine

B Lamotrigine

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Objective: Is the use of intravenous immunoglobulin in newborn infants with isoimmune haemolytic jaundice effective in reducing the need for exchange transfusion.

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Design: Systematic review of randomised and quasi-randomised controlled trials.

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Method: Seven studies were identified. Three of these fulfilled the inclusion criteria although 2 did not describe the method of allocation concealment. These 3 studies included a total of 189 infants. None of the studies used a placebo in the control group or described any method of blinding of intervention after allocation. Term and preterm infants with rhesus and ABO incompatibility were included.

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Outcome: use of exchange transfusion.

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Results: Relative risk of the use of exchange transfusion in the immunoglobulin treated group was 0.28, (95% confidence intervals (CI) 0.17 - 0.47) and numbers needed to treat (NNT) was 2.7. Weighted Mean Difference of exchange transfusions per infant in the immunoglobulin treated group was: -0.52 (95% CI -0.70 to -0.35).

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Based on the information provided above, which of the following statements is the most appropriate conclusion to draw? (5 marks)

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Select one answer only

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A NNT of 2.7 means that 2.7 exchange transfusion would have been required for each intravenous immunoglobulin administration.

B The number of exchange transfusions per infant was less in the group which did not receive immunoglobulin.

C The result of this systematic review is influenced by the lack of use of placebo in the control group.

D There was a reduction in the need for exchange transfusion in those treated with intravenous immunoglobulin.

E This systematic review provides robust evidence for the routine use of intravenous immunoglobulin for the treatment of isoimmune haemolytic jaundice.

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A 3 day old baby girl was admitted with poor feeding, and recurrent apnoeas. Examination demonstrates mild central cyanosis, a harsh systolic murmur and poor peripheral pulses. She requires resuscitation and ventilatory support. Arterial blood gases show a severe metabolic acidosis. She responds well to prostaglandin E1 infusion and oxygen saturation increased from 84% to 95%.

What is the most likely diagnosis? (4 marks)

Select one answer only

(SH 0004 SP)

A Aortic stenosis

B Endocardial cushion defect

C Pulmonary atresia

D Transposition of the great arteries with VSD

E Truncus arteriosus

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A 4 week old baby boy is seen in outpatients with a history of sudden jerky movements noticed by his mother almost from birth. They are seen most prominently whilst the baby is asleep. His mother thought they are more obvious of recent and may number from 1 or 2 to about 10 or more in a few hours sleep. Each jerk seems to last a few seconds and appears to involve an isolated limb. Physical examination is normal.

Which of the following is the most likely diagnosis? (4 marks)

Select one answer only

A Benign sleep myoclonus

B Drug withdrawal syndrome

C Hypocalcaemia

D Infantile spasms

E Myoclonic epilepsy



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A baby boy born at term developed meconium aspiration syndrome requiring ventilation. His ventilator settings were:

PIP	28 cm H ₂ O,
PEEP	5 cm H ₂ O,
rate	50/minute,
Inspiratory time	0.4 seconds
FiO ₂	1.0
Mean airway pressure	12 cm H ₂ O

His heart rate was 140/minute and oxygen saturations 82%. Mean arterial blood pressure was 45 mmHg.

Arterial blood gases:

pH	7.21	(7.35 - 7.45)
pCO ₂	5.4 kPa	(4.6 - 6.4)
pO ₂	3.2 kPa	(11 - 15)
bicarbonate	18 mmol/l	(14 - 28)
base excess	-10 mmol/l	(-2 to +2)

His chest x-ray was consistent with meconium aspiration.

Which of the following is the most appropriate next change to make in management? (4 marks)

Select one answer only

(D 0006 SP)

A Add inhaled nitric oxide at 20 ppm

B Half correct acidosis with a bicarbonate infusion

C Increase respiratory rate to 55/minute

D Start dobutamine at 5 mcg/kg/hr

E Start high frequency oscillation (HFO)



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A baby boy, birth weight 3.5 kg, was born at term in a district general hospital. The delivery involved a difficult forceps procedure following evidence of fetal distress and an abnormal CTG during labour. At birth there was no detectable heartbeat and the Apgar scores were 1 at 1 minute and 4 at 5 minutes. He was intubated and transferred to NICU. Test results are shown below.

Investigations:

Blood gases 50 minutes post delivery

pH	7.0	(7.35 - 7.45)
pCO ₂	6.9 kPa	(4.6 - 6.4)
pO ₂	11.0 kPa	(11 - 15)
bicarbonate	12.5 mmol/l	(14 - 28)
base excess	-17 mmol/l	(-2 to +2)

Pending transfer to the tertiary NICU, which of the following is the most appropriate action? (5 marks)

Select one answer only

(D 1121 SP)

- A Give bicarbonate
- B Give prophylactic phenobarbitone
- C Hyperventilate the baby to reduce intracranial pressure
- D IV normal saline 10 ml/kg bolus
- E Passive cooling

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A 15 year old boy with a diagnosis of asthma for 10 years, was referred by his GP to outpatients because of persisting respiratory symptoms. At the time of referral he was on inhaled beclometasone 1 milligram twice daily and inhaled salmeterol 50 micrograms twice daily for the last 6 months. He was also on inhaled salbutamol 2-10 puffs as required. His Peak Expiratory Flow rate was 255 L/minute (71% predicted). He was not able to participate fully in school sports.

Which two of the following investigations should you do next? (8 marks)

Select two answers only

A Bronchoscopy and bronchial biopsy

B Mantoux test

C Nasal brushings

D Serum IgE and RAST tests to aeroallergens

E Short synacthen test

F Spirometry with reversibility

G Spiral CT of chest

H Sweat test

I Ventilation/perfusion lung scan



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A previously well 16 year old boy presented to outpatients with a 6 month history of frequent headaches. Recent problems with balance had occurred while playing football for his school team. The headaches were relieved by lying flat. There was a strong family history of migraine.

Physical examination is normal apart from a positive Romberg sign and unsteadiness on attempting to heel-toe walk. Reflexes are normal.

Which of the following is the most likely diagnosis? (4 marks)

Select one answer only

A Arnold-Chiari malformation type 1

B Idiopathic (benign) intracranial hypertension

C Late onset muscular dystrophy (Becker)

D Migraine

E Spinal cord tumour



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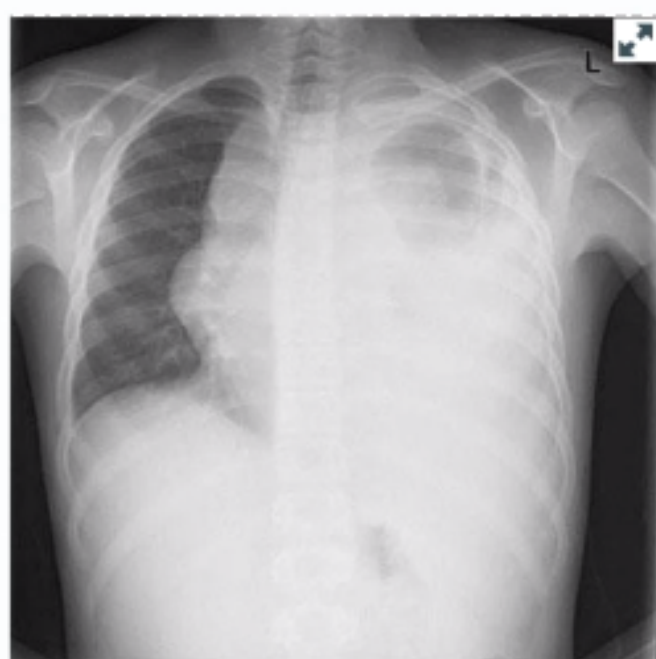
A 13 year old boy was referred by his GP with a 5 day history of cough and increasing breathlessness. His chest x-ray is shown below.

What are the two radiological abnormalities seen on the x-ray? (6 marks)

Select two answers only

Clicking on the x-ray file below will open it in a new window, allowing it to be moved around the screen

(X 3071)



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A 12 year old girl presented to the emergency department with severe abdominal pain and rectal bleeding. She was observed to have 2 episodes of bilious vomiting.

She was previously seen in clinic with recurrent episodes of crampy abdominal pain lasting up to an hour and associated with pallor. Investigations at that review, including a coeliac screen and abdominal ultrasound, were normal.

On examination, her heart rate is 140/minute, capillary refill time 3 seconds, she is afebrile, a small, slightly tender mass is palpable in the right iliac fossa and bowel sounds are absent.

Which of the following is the most likely explanation for her symptoms? (4 marks)

Select one answer only

A Anal fissure

B Campylobacter enteritis

C Crohn's colitis

D Henoch-Schönlein purpura

E Intussusception

F Volvulus



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A 12 year old boy with type 1 diabetes since the age of 8 comes to clinic for his annual review. His growth is normal for age, his control has been excellent and his HbA1c is 42 mmol/mol (6%).

Which two of the following conditions known to be associated with type 1 diabetes should be screened for at this age? (6 marks)

Select two answers only

(SH 0014 SP)

- A Coeliac disease
- B Exocrine pancreatic insufficiency
- C Hypercholesterolaemia
- D Nephropathy
- E Peripheral neuropathy
- F Retinopathy

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A 2 year old African Caribbean girl was referred with concern about her locomotor development. Further history taking revealed that she was largely breast-fed and was described as a "fussy eater". She lived with her mother and new boyfriend in a high rise block of flats. Her height and weight are on the 2nd to 9th centiles for age. Her wrist x-ray is shown below.

Which of the following is the most likely explanation for her symptoms? (4 marks)

Select one answer only

Clicking on the x-ray image below will open it up in a new window, allowing it to be moved around the screen



A Lead poisoning

B Mucopolysaccharidosis

C Osteogenesis imperfecta

D Osteopetrosis

E Rickets

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A 12 year old boy had been diagnosed with Hodgkin lymphoma 2 months ago. His staging investigations showed that he had disease localised to the mediastinum. Chemotherapy was started following the international protocol with doxorubicin, bleomycin, vinblastine and dacarbazine (ABVD) chemotherapy every 3 weeks for a planned 6 month duration. He had managed to return to school for a few hours on a daily basis. He was then re-admitted with a 2 day history of dry cough and temperature.

On examination, he is pale and with some difficulty in speaking. His temperature is 38.2°C and his oxygen saturation is 88% in air. His heart rate is 100, BP 100/60 and heart sounds are normal. He has nasal flaring, is tachypnoeic at rest with a rate of 40 breaths per minute and has bilateral intercostal recession. Auscultation reveals normal breath sounds with the occasional crepitation. Supplemental oxygen is started, IV antibiotics provided and a CXR is requested.

Which of the following is the most likely explanation for this clinical presentation? (4 marks)

Select one answer only

A Bleomycin induced pulmonary toxicity

B Influenza A pneumonia

C Pneumococcal pneumonia

D Pneumocystis jirovecii pneumonitis

E Relapse of mediastinal Hodgkin lymphoma

F Respiratory Syncytial Virus pneumonitis

Applied Knowledge in Practice – Sample Paper 1

Answer Key

Question 1

Question No: (C 1691 SP)

Answer: *B - Eczema herpeticum*

Reasoning:

The child has widespread vesiculo-pustular lesions involving the face and upper trunk which is typical of eczema herpeticum – atopic dermatitis infected with herpes simplex. The history of a mild skin rash since the age of two months suggests underlying chronic condition - eczema.

Answer A is wrong

The rash of chicken pox evolves from pruritic, erythematous macules to papules and then vesicles. The rash occurs in crops, so it is characteristic to find lesions at various stages. There is usually relative sparing of the face in chickenpox. Varicella in an immune-compromised child may cause this appearance, but it is much less likely than eczema herpeticum, particularly with the history that he had been previously well.

Answer B is the best answer

Answer C is wrong

Impetigo usually starts with a vesicle or pustule which develops into a crusted, oozing lesion which then gradually spread.

Answer D is wrong

Primary herpes simplex infection presents with gingivo-stomatitis. Vesicles develop in the mouth and on the tongue and lips, but would not involve the rest of the face and the upper trunk.

Answer E is wrong

Stevens-Johnson syndrome starts with erythematous macules with central necrosis. The lesions then form vesicles, bullae and desquamation. The condition involves two or more mucosal surfaces that may include the eyes, oral cavity, upper airway and G-I tract.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXXI. The Skin.

Question 2

Question No: (X 0020 SP)

Answer: *C - Congenital lung cyst*

Reasoning:

The obvious abnormality on this chest x-ray is the round, air-filled lesion in the right lower lobe. This child is basically well with a cough which will exclude those answers leading to significant lung compromise.

Answer A is wrong

There are no bowel gas patterns within either lung field.

Answer B is wrong.

Congenital lobar emphysema (or congenital lobar hyperinflation) generally affects the upper lobe, or right middle lobe. The hyperlucency would be in a lobar pattern. This is in the lower lobe area and not lobar. The child would be unwell with a tachypnoea.

Answer C is the best answer

A localised air-filled lesion in a well-child would explain the history and findings.

Answer D is wrong

The child would be unwell with a tachypnoea. A loculated pneumothorax would be unlikely, as it is normally loculated due to pleural adhesions and would be unlikely to persist for a month.

Answer E is wrong.

A pneumatocoele is seen in Staphylococcal lung infection and the patient would be extremely unwell

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XIX The Respiratory System

Question 3a

Question No: (EMQ 0007 SP a)

Answer: *A - Bronchiectasis*

Reasoning:

The child has a persistent cough and focal signs after a lower respiratory infection.

Acute infections such as chlamydia and pertussis might have caused the original illness, but not the sequelae.

There is no history given to suggest TOF, aspiration, gastro-oesophageal reflux, hyper IgE or tuberculosis. Obliterative bronchiolitis is a potential consequence of LRTI and can be associated with bronchiectasis but would normally have associated wheeze with fixed lower airway obstruction.

PCD is a plausible cause of chronic chest symptoms and signs, but would normally have presented with earlier onset of symptoms, and associated ear and nose problems, rather than a single LRTI.

Thus, bronchiectasis is the best explanation of these symptoms and signs.

Further Reading

Up to Date: <https://www.uptodate.com/contents/epidemiology-clinical-presentation-and-evaluation-of-parapneumonic-effusion-and-empyema-in-children> Fakhoury K; Kanu A
[Accessed May 2018]

Question 3b

Question No: (EMQ 0007 SP b)

Answer: *I - Recurrent aspiration*

Reasoning:

Recurrent cough and wheeze indicates a chronic condition.

It is likely that a child who suffered from severe birth asphyxia has neurodevelopmental consequences, of which the commonest is cerebral palsy. The likeliest cause of respiratory symptoms in a child with cerebral palsy is aspiration, either from gastro-oesophageal reflux or pharyngeal incoordination.

Thus, recurrent aspiration is the most accurate answer to the question.

Further Reading

Up to Date: <https://www.uptodate.com/contents/management-and-prognosis-of-cerebral-palsy>. Author: Patterson MC; [Accessed May 2018]

Question 3c

Question No: (EMQ 0007 SP c)

Answer: *D - Hyper IgE syndrome*

Reasoning:

The key features in this case are the recurrent skin sepsis and recurrent cough and sputum, colonised with *Staphylococci aureus*. *Staphylococci* are unusual pathogens in sputum, and two conditions are suggested by this - cystic fibrosis and Hyper-IgE syndrome.

The former is not in the list, and the latter is associated with skin sepsis because of the specific immune deficiency.

Further Reading

Up to Date <https://www.uptodate.com/contents/autosomal-dominant-hyperimmunoglobulin-e-syndrome> Authors: LaPine A; Kumánovics TR; and Hill HR; [Accessed May 2018]

Question 4

Question No: (EH 39 SP)

Answer: *A - Anti-double stranded DNA*

Reasoning:

The photograph shows a young person with a facial rash across both malar regions and the bridge of the nose – a ‘butterfly distribution’. The history suggests sensitivity to sunlight, chest and joint symptoms along with episodic pyrexia. A likely chronic anaemia and raised CRP are seen. All these observations suggest a diagnosis of Systemic Lupus Erythematosus (SLE) - in this case exacerbated by UV light (excess sunlight) exposure.

Answer A is the best answer

A positive result for anti-double stranded DNA will confirm the diagnosis of SLE

Answer B is wrong

Although temperatures, chest signs and arthropathy can be seen in Mycoplasma infection, the rash is not seen.

Answer C is wrong

Parvovirus gives the ‘slapped cheek’ facial rash but the illness is usually short lived. It has been associated with arthropathy but this is not a common feature.

Answer D is wrong

Systemic symptoms are seen in rheumatoid disease along with similar abnormalities shown in the blood results but the rash is more characteristic of SLE.

Answer E is wrong

The symmetrical distribution of the rash is not characteristic of a superficial infection.

Answer F is wrong

Porphyria can lead to a photosensitivity of the skin but would not produce the marked arthropathy described here

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XVI Rheumatic Diseases of Childhood.

Question 5

Question No: (SH 2278 SP)

Answer: *B - Blood sugar*

Answer: *C - Calcium*

Reasoning:

All of these answers are plausible and indeed all of them might be requested. But the question asks which two would be most helpful in his immediate management.

Hypoglycaemic seizures in infancy are associated with poor neurological outcome so it is crucial to check the glucose as the infant arrives and correct it as necessary.

Similarly, hypocalcaemia is easily and quickly diagnosed and must be corrected. 'Treat the Treatable' is a good adage.

Answer A is wrong

Although the history may be consistent with a urea cycle defect, his mental state does not suggest this, and the condition would not be addressed immediately at presentation

Answer B is the best answer

Crucial investigation that allows immediate correction

Answer C is the best answer

Crucial investigation that allows immediate correction

Answer D is wrong

There is no history of a bleeding disorder

Answer E is wrong

May be an appropriate investigation for the presentation but request, implementation and interpretation would not be immediate.

Answer F is wrong

May be an appropriate investigation for the presentation but request, implementation and interpretation would not be immediate.

Answer G is wrong

Anaemia and thrombocytopaenia may be causes for the presentation but there is no history of pallor or bruising.

Answer H is wrong

Abnormalities of sodium, potassium, creatinine and urea would be important to know and abnormal sodium could lead to seizure activity but are less likely than hypocalcaemia in this child.

Further Reading

UpToDate <https://www.uptodate.com/contents/etiology-and-prognosis-of-neonatal-seizures>

Author: Shellhass R. [Accessed May 2018]

Question 6

Question No: (C 2064 SP)

Answer: *D - Meckel's diverticulum*

Reasoning:

This is an unusual scenario but the history is highly suggestive of pathology.

Blood mixed with the stools suggests lower gastro-intestinal bleeding and merits investigation. This is a radio-isotope scan that is used to make a diagnosis of a Meckel's diverticulum – the injected material is taken up by the mucosal cells found in stomach (or elsewhere). The image shown is of the abdominal area of the child and reveals the expected uptake of the stomach mucosa (dense area at the top right), the contrast in the bladder (at the bottom middle of the picture) and the ectopic mucosa of the Meckel's diverticulum (in the centre of the scan).

Answer A is wrong

The child with an appendix abscess would have a short history and would likely be toxic. The 99mTc pertechnetate scan would not be used in such a situation.

Answer B is wrong

The child with an intussusception would have a short history and would likely be acutely unwell. The 99mTc pertechnetate scan would not be used in such a situation.

Answer C is wrong

The child with a malrotation would have a short history and would likely be acutely unwell. The 99mTc pertechnetate scan would not be used in such a situation.

Answer D is the best answer

Answer E is wrong

The history is not that of vesico-ureteric reflux and a MAG3 scan would be the more appropriate investigation.

Further Reading

UpToDate: <https://www.uptodate.com/contents/lower-gastrointestinal-bleeding-in-children-causes-and-diagnostic-approach> Authors: Patel N; Kay M. [Accessed May 2018]

Question 7

Question No: (C 0005 SP)

Answer: *E - Intraocular pressure measurement*

Reasoning:

This is a two-stage question. The first stage is to make a diagnosis. The unilateral capillary haemangioma (port wine stain) is typical of the Sturge-Weber syndrome.

The second stage is to consider complications of this syndrome that may occur in infancy and consider an appropriate investigation for that condition.

Answer A is wrong

Sturge-Weber syndrome is not genetically determined and therefore chromosome analysis is irrelevant.

Answer B is wrong

An MRI scan with gadolinium is important to look for an associated intracranial vascular abnormality, but there is no immediate urgency for this

Answer C wrong

Cranial ultrasound will not show a vascular abnormality and is not indicated

Answer D is wrong

Although around 80% of children with Sturge-Weber Syndrome develop epilepsy an EEG is not indicated until the child presents with a seizure.

Answer E is the best answer

Glaucoma affecting the ipsilateral eye is a common complication in infancy and appropriate assessment is mandatory

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXXI. The Skin.

Question 8

Question No: (D 0003 SP)

Answer: C - HIV/AIDs

Reasoning:

This child has recently arrived in the UK and so past medical history is not known. The child may come from an area where some conditions are more prevalent. This child has poor feeding, loose stools and faltering growth. The child is also anaemic and has raised immunoglobulins.

Answer A is wrong

The child with chronic granulomatous disease has neutrophil dysfunction and therefore usually presents with recurrent and multiple abscesses

Answer B is wrong

Common Variable Immune Deficiency is characterised by hypogammaglobinaemia and that is not demonstrated by the immunoglobulin results.

Answer C is the best answer

HIV / AIDS presents with failure to thrive, chronic diarrhoea, chronic parotid swelling, lymphocytic interstitial pneumonitis (LIP) and oral thrush.

Answer D is wrong

In Severe Combined Immune deficiency, the lymphocytes will be low – often less than $0.5 \times 10^9/l$ - and immunoglobulin levels are reduced.

Answer E is wrong

In X-linked hypogammaglobinaemia the immunoglobulins will be low

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XVII Infectious Diseases

Question 9

Question No: (C 0007 SP)

Answer: *D - Staphylococcal scalded skin syndrome*

Reasoning:

The child has diffuse erythema of the face with peeling of the skin periorally, around the eyes and in the neck. The epidermis has separated from the dermis to form blisters that have then ruptured. The folds of lifted epidermis can be seen (above both eyes, left cheek, mouth, chest). This feature is typical of staphylococcal scalded skin syndrome and pemphigus.

Answer A is wrong

Atopic dermatitis in infancy characteristically involves the cheeks and scalp. The skin is erythematous with dry crusting lesions. Perioral involvement and desquamation are not features of this condition and the unaffected scalp is also against this diagnosis.

Answer B is wrong

Herpes simplex at this age is usually a primary infection that presents with gingivo-stomatitis. Vesicles develop in the mouth and on the tongue and lips.

Answer C is wrong

Impetigo usually starts with a vesicle or pustule which develops into a crusted, oozing lesion which then gradually spread.

Answer D is the best answer

Answer E is wrong

Stevens-Johnson syndrome starts with erythematous macules with central necrosis. The lesions then form vesicles, bullae and desquamation. The condition involves two or more mucosal surfaces which may include the eyes, oral cavity upper airway and G-I tract.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XVII Infectious Diseases

Question 10

Question No: (X 0011 SP)

Answer: *B - Fluid resuscitation with 10 ml/kg 0.9% saline*

Reasoning:

The clinical story is that of a preterm infant who is in circulatory shock -tachycardia, hypotension and peripheral oedema. His blood gas shows a profound metabolic acidosis. Abdominal x-ray shows dilated loops of bowel, thickening of the bowel wall and free air at the very top of the film – features of NEC and perforation. Resuscitation is required with urgency. The question asks for the ‘most appropriate immediate management’; although more than one action may be necessary in clinical management; the question is asking you to identify the response with the greatest urgency.

Answer A is wrong

Although the baby has a metabolic acidosis, the cause of this is primarily due to circulatory shock. Bicarbonate would not address the cause of the metabolic problem.

Answer B is the best answer

Answer C is wrong

Increasing ventilatory pressures and rates would drop the CO₂ but this is currently within the normal range and the action would again not address the causative problem.

Answer D is wrong

No place in the described situation.

Answer E is wrong

These would be needed promptly but they would not be the ‘most appropriate immediate management’

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XII. The Fetus and Neonatal Infant

Question 11

Question No: (7000 SP)

Answer: *G - Reflex anoxic seizures*

Reasoning:

Answer A is wrong

The episodes do not occur at night

Answer B is wrong

The history does not describe seizures with abnormal, coordinated actions

Answer C is wrong

There is nothing in the given history that raises this as a diagnosis and probably there were independent witnesses to the event at the nursery.

Answer D is wrong

Child has a 'mild elevation of temperature' prior to one these episodes but that is unlikely to cause seizure activity.

Answer E is wrong

No record of glucose results so makes this explanation unlikely.

Answer F is wrong

The episodes are described as 'shaking' and not 'jerking'

Answer G is the best answer

Classical history where pain or anger initiates a 'crying response' and the child collapses often prior to the onset of the cry. Benign, self-limiting and resolve in time.

Further Reading

<http://www.childneurologyfoundation.org/disorders/breath-holding-spells/> [Accessed May 2018]

Question 12 a

Question No: (C 0011 SP a)

Answer: *E - Inadequate calorie intake*

Reasoning:

The growth chart shows you that the head circumference follows the 50th centile and the most recent measurement of length is also on the 50th. The weight of the child follows the 50th centile until 20 weeks then falls to the 0.4 centile. The child is admitted to hospital and the weight moves up to the 9th centile. The stem to the question does not indicate that any specific intervention or treatment given – therefore assume that no active treatment was provided. It is clear that the hospital admission confirms good weight gain can be achieved when adequate calorific intake is assured.

Answer A is wrong

Although the start of poor weight gain coincided with the introduction of solids, a child with coeliac disease would not usually present with vomiting and any weight gain would have been the result of a move to a gluten-free diet (not mentioned). The weight gain would not have been so dramatic.

Answer B is wrong

A child with CMPI would usually have diarrhoea along with the vomiting and a change in diet would have been mentioned in the text.

Answer C is wrong

If the child had cystic fibrosis then the observed increase in weight would have been the result of an intervention – and this would have been mentioned in the text.

Answer D is wrong

If the child had GORD then the observed increase in weight would have been the result of an intervention – and this would have been mentioned in the text.

Answer E is the best answer

This child has therefore not been given sufficient calories for weight gain and neglect is the most likely cause.

Further Reading

Clinical Cases for MRCPCH Applied Knowledge in Practice RCPCH 2016. Ed R Dinwiddie.

Question 12b

Question No: (C 0011 SP b)

Answer: *A - Discharge planning meeting*

Reasoning:

The most appropriate answer to this second part of the question depends upon recognising the appropriate diagnosis from part 1.

Answer A is the best answer

The chart shows you that the most likely explanation is neglect with inadequate calories provided. A full assessment by all professionals involved – hospital staff, community team and social workers is needed to assess the potential risk to the child if discharged. The first concern has to be the safety of the child.

Answer B is wrong

Inappropriate investigation where safeguarding is the recognise cause of the problem.

Answer C is wrong

Inappropriate investigation where safeguarding is the recognise cause of the problem.

Answer D is wrong

Inappropriate investigation where safeguarding is the recognise cause of the problem.

Answer E is wrong

Inappropriate investigation where safeguarding is the recognise cause of the problem.

Further Reading

Clinical Cases for MRCPCH Applied Knowledge in Practice RCPCH 2016. Ed R Dinwiddie.

Question 13a

Question No: (EMQ 0003 SP a)

Answer: *B - Lamotrigine*

Reasoning:

First line treatment of Juvenile absence epilepsy should be with one of the following - ethosuximide, sodium valproate or lamotrigine. A girl of this age - who may become pregnant - should not be offered sodium valproate due to its teratogenic effect.

Ethosuxamide is not on the list of options and so lamotrigine is the most appropriate treatment.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXVII. The Nervous System

Question 13b

Question No: (EMQ 0003 SP b)

Answer: *I - Vigabatrin*

Reasoning:

The first line option in infantile spasms would either be ACTH or vigabatrin. ACTH is not an option offered here,

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXVII. The Nervous System

Question 13c

Question No: (EMQ 0003 SP c)

Answer: *D - No treatment*

Reasoning:

The patient has benign childhood epilepsy with centrotemporal spikes.

The option of not treating the child should be considered particularly if not impacting on lifestyle. The seizures are often at night and are infrequent in nature. Status epilepticus is very uncommon. The prognosis is very good and seizures will resolve in most children often by the early teenage years.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXVII. The Nervous System

Question 14

Question No: (EBM 0013 SP)

Answer: *D - There was a reduction in the need for exchange transfusion in those treated with intravenous immunoglobulin.*

Reasoning:

The “95% confidence interval” means that we are 95% confident that the population value of interest lies within this given range.

The “relative risk” indicates the risk of an event occurring in an exposed group when compared with the risk of the event occurring in an unexposed group. A “relative risk” (expressed as a ratio) around 1 indicates little or no difference, greater than 1 indicates a greater risk of the event happening in the exposed group and a value less than 1 indicates a reduced risk of the event happening in the exposed group.

Answer A is wrong

The concept of ‘Numbers Needed to Treat’ is a measure of how many individuals need to be treated for one individual to benefit. In this study for every 2.7 individuals treated, 1 patient will have a beneficial result with immunoglobulins when compared with the control group. It does not refer to the number of transfusions.

Answer B is wrong

This statement is the opposite of what was found. The number of exchange transfusions per infant was less in the group that received immunoglobulin.

Answer C is wrong

A systematic review identifies those published research studies which meet a predefined set of criteria and thereby allow specific questions to be addressed. If a study meets the criteria then it is included. The design of the study cannot then be challenged if it meets these criteria.

Answer D is the best answer

There was a reduction in the need for exchange transfusion in those treated with intravenous immunoglobulin as the weighted mean was a negative value [-0.52] and the confidence interval range was negative – i.e. fewer exchanges needed.

Answer E is wrong

While the review is robust, evidence of the effect of intravenous immunoglobulins on the need for exchange transfusion, the introduction of this therapy as routine would require a careful risk-benefit assessment, which cannot be made on the data available.

Further Reading

R Gottstein, R W I Cooke Systematic review of intravenous immunoglobulin in haemolytic disease of the newborn Arch Dis Child Fetal Neonatal Ed 2003;88:F6–F10

Question 15

Question No: (SH 0004 SP)

Answer: *D - Transposition of the great arteries with VSD*

Reasoning:

Answer A is wrong

Aortic stenosis could produce the symptoms recorded but would not produce the observed central cyanosis. Improvement in the oxygenation with the prostaglandin can be seen and this would not occur in aortic stenosis.

Answer B is wrong

AVSD would produce increased pulmonary blood flow and possibly heart failure but do not produce cyanosis and would not improve with prostaglandin

Answer C is wrong

Babies with pulmonary atresia become acutely unwell as the duct closes within the first few hours. Prostaglandin E₁ infusion is necessary and early surgical intervention required. The child would not present at 3 days of age.

Answer D is the best answer

A baby with TGA and VSD has some mixing of oxygenated and deoxygenated blood in the ventricles depending on the size of the VSD. If the VSD is small then the lesion is duct dependent but may present later.

Answer E is wrong

A combined outflow tract from both left and right ventricles allows mixing of bloods and is unlikely to present with acidosis. The lesion is not duct dependent, so would not improve with prostaglandin.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 16

Question No: (SH 0016 SP)

Answer: A - *Benign sleep myoclonus*

Reasoning:

The diagnosis of the cause of 'funny turns' or abnormal movements in any child relies predominantly on the history. Here we have jerking movements seen only when the child sleeps. They are of short duration.

Answer A is the best answer

This is not the history of pathological seizures of any type – the episodes are limited to the time the baby is asleep. Benign sleep myoclonus becomes the most likely diagnosis by the exclusion of the other offered diagnoses. Benign sleep myoclonus is a common and self-limiting condition.

Answer B is wrong

Symptoms from drug withdrawal are not confined to sleep and would not be increasing in frequency at this age.

Answer C is wrong

Hypocalcaemia characteristically causes 'jitteriness' but, if severe, can lead to generalised seizures. Symptoms from hypocalcaemia are not confined to times of sleep.

Answer D is wrong

Symptoms from infantile spasms are not confined to sleep.

Answer E is wrong

Symptoms from myoclonic epilepsy are not confined to sleep.

Further Reading

UpToDate <https://www.uptodate.com/contents/non epileptic-paroxysmal-disorders-in-infancy> Authors: Nguyen TT; Kaplan PW; Wilfong A; [Accessed May 2018]

Question 17

Question No: (D 0006 SP)

Answer: *A - Add inhaled nitric oxide at 20 ppm*

Reasoning:

This baby has persistent pulmonary hypertension of the newborn (PPHN) as indicated by the normal CO₂ and low oxygenation. This is often seen in term babies with meconium aspiration. Pulmonary vasodilatation or ECMO may be necessary.

Answer A is the best answer

The addition of inhaled nitrous oxide would lead to pulmonary vasodilation and thereby improve arterial oxygenation.

Answer B is wrong

Blood gas bicarbonate level is at the lower limit of normal and does not need correction at this stage.

Answer C is wrong

This would drop the pCO₂ further and would not be appropriate

Answer D is wrong

The mean arterial blood pressure is currently at an acceptable level and so does not need immediate support.

Answer E is wrong

High frequency oscillation is a reasonable option and is used in the management of PPHN but is usually considered when PIP reach 28-30. The evidence of efficacy of HFOV in PPHN remains limited.

Further Reading

UpToDate <https://www.uptodate.com/contents/persistent-pulmonary-hypertension-of-the-newborn> Authors: Stark A; Eichenwald E. [Accessed May 2018]

Question 18

Question No: (D 1121 SP)

Answer: *E - Passive cooling*

Reasoning:

Answer A is wrong

This intervention is not evidence-based

Answer B is wrong

This intervention is not evidence-based

Answer C is wrong

This intervention is not evidence-based

Answer D is wrong

This intervention is not evidence-based

Answer E is the best answer

This intervention has been shown to affect outcome in severe asphyxia. This baby fulfils the cooling criteria from the UK TOBY study register.

Further Reading

Anzopardi DV et al Moderate hypothermia to treat perinatal asphyxial encephalopathy. NEJM 2009;361(14) 1349-58

Strohm, B et al; Temperature control during therapeutic moderate whole-body hypothermia for neonatal encephalopathy. Arch Dis Child Fetal Neonatal Ed 2010; 9(1):39-45

TOBY trial protocols:

<https://www.npeu.ox.ac.uk/toby/protocol> [Accessed May 2018]

Question 19

Question No: (SH 0008 SP)

Answer: *E - Short synacthen test*

Answer: *F - Spirometry with reversibility*

Reasoning:

The most common cause of poorly controlled asthma is poor adherence to therapy, especially in a teenager. Other diagnoses also need to be excluded.

Answer A is wrong

This test would be inappropriate until more basic assessments had been carried out.

Answer B is wrong

Nothing in the history suggests a diagnosis of tuberculosis

Answer C is the wrong

Ciliary dyskinesia may be considered at a later stage.

Answer D is the wrong

These may be considered at a later stage in the management.

Answer E is the best answer

If he is taking treatment as prescribed (high doses), then he is likely to have a degree of adrenal suppression, which could be dangerous.

Answer F is the best answer

A more detailed spirometric assessment would give more information than the crude measure of PEF. If obstruction is confirmed on the flow-volume loop, and reversibility is demonstrated then the diagnosis of asthma is strengthened, and the likelihood of non-adherence increases.

Answer G is wrong

CT chest is not generally felt to be worth the radiation exposure.

Answer H is wrong

Cystic fibrosis should be considered but sweat test would not be the next investigation

Answer I is wrong

Nothing in the history suggests pulmonary emboli.

Further Reading

International ERS/ATS guidelines on definition, evaluation and treatment of severe asthma..

Chung et al. European Respiratory Journal 2013; DOI: 10.1183/09031936.00202013.

[Accessed May 2018]

Question 20

Question No: (SH 0013 SP)

Answer: *A - Arnold-Chiari malformation type 1*

Reasoning:

Headaches are a common problem presenting in out-patients and one that depends heavily on a clear history. Certain 'red flag' features can indicate significant pathology. This young person has a relatively long history with some rather strange aspects – frequent, improve when lying down and added description of balance problems with exercise. The neurological examination identifies abnormalities.

Answer A is the best answer

The Arnold-Chiari malformation type 1 is a herniation of the cerebellar tonsils through the foramen magnum. [Arnold-Chiari malformation type 2 are often associated with spinal dysraphism and Arnold-Chiari malformation type 3 is extremely rare].

Answer B is wrong

Although Idiopathic (benign) intracranial hypertension does lead to chronic headaches of a persistent nature, they are not associated with abnormal neurological findings.

Answer C is wrong

A Becker muscular dystrophy leads to features of myopathy that would mainly include weakness of lower limbs.

Answer D is wrong Although there is a strong family history of migraine, this is not the pattern of headaches seen in classical migraine – these usually with episodic headaches with longer, symptom-free, periods. One would not expect abnormal neurology on examination.

Answer E is wrong

A spinal cord tumour would not give a history of headaches.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXVII. The Nervous System.

Question 21

Question No: (X 3071)

Answer: *D - Hilar lymphadenopathy*

Answer: *F - Left pleural effusion*

Reasoning:

This chest x-ray is grossly abnormal and shows dense shadowing in the mediastinum and the left side of the chest.

The large left pleural effusion and gross bilateral hilar lymphadenopathy are likely the result of some form of non-Hodgkin lymphoma or leukaemia and those which arise in the chest are usually T-cell in origin (from thymic (T) modulated cells).

Answer A is wrong

Cardiac shadow is visible behind the shadowing and is normal

Answer B is wrong

Cardiac shadow is visible and is normal

Answer C is wrong

No evidence of fluid in the right horizontal fissure.

Answer D is the best answer

Appearances are characteristic of bilateral hilar lymphadenopathy. This is less clear on the left because of the dense shadowing (fluid) occupying the left hemithorax.

Answer E is wrong

No evidence of a left Gohn focus.

Answer F is the best answer

There is a fluid level extending up the lateral chest wall consistent with a large left pleural effusion and this causes some midline shift to the right.

Answer G is wrong

There is some tracheal shift to the right but no air in the pleural cavity on the left.

Answer H is wrong

The upper part of the central shadow is consistent with thymic enlargement but the mass is much larger than could be attributed to just thymus (persistent thymic shadow)

Answer I is wrong

The shape of the cardiac shadow is not consistent with this diagnosis.

Further Reading

UpToDate <https://www.uptodate.com/contents/epidemiology-clinical-presentation-and-evaluation-of-parapneumonic-effusion-and-empyema-in-children> Authors: Janahi I ; Fakhoury K [Accessed May 2018]

UpToDate <https://www.uptodate.com/contents/clinical-assessment-of-the-child-with-suspected-cancer> Authors: Neville K; Steuber CP [Accessed May 2018]

Question 22

Question No: (7001 SP)

Answer: *E - Intussusception*

Reasoning:

This girl has an acute abdomen with evidence of shock. In this context the rectal bleeding is suggestive of ischaemic bowel.

In this list the only 2 possibilities causing this are volvulus and intussusception.

The palpable mass and the rectal bleeding makes intussusception most likely.

Although intussusception is most common in infants and toddlers, about 10% of cases occur in children over five years and 3 -4 % in those over 10 years; and 1% in infants <3 months. Outside the typical age range, it is likely to be associated with a pathological 'lead' point, which may include reactive lymphoid hyperplasia, small bowel lymphoma or a Meckel's diverticulum.

Further Reading

UpToDate: <https://www.uptodate.com/contents/lower-gastrointestinal-bleeding-in-children-causes-and-diagnostic-approach> Authors: Patel N; Kay M; [Accessed May 2018]

<https://www.uptodate.com/contents/intussusception-in-children> Authors: Vo N; Sato TT [Accessed May 2018]

Question 23

Question No: (SH 0014 SP)

Answer: *D - Nephropathy*

Answer: *F - Retinopathy*

Reasoning:

Answer A is wrong

Would not form part of an annual assessment, although offered at diagnosis..

Answer B is wrong

Would not form part of an annual assessment.

Answer C is wrong

Would not form part of an annual assessment.

Answer D is the best answer

Vasculopathy is a significant issue and will affect renal tissue. Annual check for albuminuria is vital.

Answer E is wrong

Peripheral neuropathy is a recognised complication of type 1 diabetes but would not be common at this age.

Answer F is the best answer

Vasculopathy leads to a diabetic retinopathy and is more likely with duration of diagnosis and degree of glycaemic control.

Further Reading

UpToDate <https://www.uptodate.com/contents/complications-and-screening-in-children-and-adolescents-with-type-1-diabetes-mellitus> Authors: Levitsky, L; Misra, M; [Accessed May 2018]

Question 24

Question No: (X 0002 SP)

Answer: *E - Rickets*

Reasoning:

The X-ray of the wrist shows poor mineralisation associated with cupping and fraying of the metaphyseal region.

Answer A is wrong

Lead poisoning gives dense lines along the metaphyseal margin (look for a picture)

Answer B is wrong

Mucopolysaccharidosis is a collection of conditions caused by enzyme deficiencies leading to abnormal storage of glycosamines in connective tissue. The radiological changes are extensive and include short, thick phalanges with proximal widening.

Answer C is wrong

The radiological appearances of osteogenesis imperfecta are osteopenia and fractures.

Answer D is wrong

The bones are dense white in osteopetrosis.

Answer E is the best answer

This is classical of Vitamin D deficiency rickets (poor mineralisation associated with cupping and fraying), which occurs in a baby who is largely breast fed, fussy eater and who is failing to thrive.

Further Reading

<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3731470/> [Accessed May 2018]

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XI. Metabolic Disorders.

Question 25

Question No: (7002 SP)

Answer: D - *Pneumocystis jirovecii pneumonitis*

Reasoning:

Answer A is wrong

Bleomycin does cause a fibrosis and could present with some of the features listed here but not so acutely or so profoundly and the patient would not be pyrexial.

Answer B is wrong

Influenza A pneumonia would not be expected to give such marked hypoxia in the early phases

Answer C is wrong

Pneumococcal pneumonia that was the cause of such marked hypoxia would produce a toxic presentation in the patient with much higher temperatures

Answer D is the best answer

Pneumocystis jirovecii pneumonitis is seen in patients who are immunosuppressed and they will often have marked hypoxia despite a paucity of chest findings on auscultation.

Answer E is wrong

Relapse of mediastinal Hodgkin lymphoma may cause some features such as pyrexia but individuals would usually present with symptoms long before they developed such hypoxia.

Answer F is wrong

Respiratory Syncytial Virus pneumonitis would not be expected to give such marked hypoxia in the early phases

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XIX The Respiratory System

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This is the tongue of an otherwise healthy child. Her mother is worried about the appearance.

Which of the following is the most appropriate management? (4 marks)

Select one answer only

Clicking on the image below will open it in a new window, allowing it to be moved around the screen

(C 0012 SP)



A Biopsy of the white area

B Determine HIV status

C Follow up for observation

D Nystatin

E Offer reassurance



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A 3 day old girl presented with swelling and bruising of the right eyelid. She had been born at term by ventouse delivery due to fetal distress. Birth weight was 2.3 kg and she did not require resuscitation. She vomited 15 minutes after receiving oral vitamin K. She was otherwise well and feeding had been established without difficulty. Her test results are shown below.

Investigations:

Blood

haemoglobin	144 g/l	(145 - 220)
white cell count	6.5 x 10 ⁹ /l	(10.0 - 26.0)
normal differential		
platelets	29 x10 ⁹ /l	(150 - 450)
CRP	5 mg/l	(< 5)

Which of the following is the most important information contributing to the explanation of her problem? (5 marks)

Select one answer only

(D 0009 SP)

A Family history of easy bruising

B Low maternal platelet count in pregnancy

C Maternal anti-epileptic medication during pregnancy

D Perinatal risk factors for Group B Streptococcal sepsis

E Vomiting after vitamin K administration

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Title: Short versus long duration of antibiotic therapy for bacterial meningitis: a meta-analysis of randomised controlled trials in children.

Objective: To evaluate the effectiveness of short courses (<7 days) versus long courses (7-14 days) of antibiotic therapy for bacterial meningitis.

Outcomes: Clinical success, mortality, duration of hospitalisation, adverse events, hearing impairment, long-term neurological complications.

Results: Five open-label RCTs involving a total of 367 patients were included. The odds ratios for short versus long course outcomes were as follows: long term neurological complications 0.60 (95% confidence interval (CI) 0.29-1.27), hearing impairment 0.59 (95% CI 0.28-1.23). The weighted mean difference in the duration of hospitalisation was -2.17 days (95% CI -3.85 to -0.50).

(Adapted from ADC 2009;94:607-614)

Which two of the following conclusions can be deduced from the data? (6 marks)

Select two answers only

A Long course treatment resulted in higher levels of hearing impairment.

B Meta-analysis of RCTs is not the most appropriate way to resolve such clinical issues.

C Patients treated for meningitis should be observed in hospital for 24 hours following cessation of treatment.

D Short course treatment resulted in a significantly reduced length of hospital stay.

E Short course treatment should become the standard treatment for bacterial meningitis in children.

F There was no difference in the level of neurological complications between treatment groups.



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The mother of an 8 year old girl with type 1 diabetes reports that her daughter is having shaking episodes during her sleep between 6 and 7 am. During the episodes, the girl is unresponsive and afterwards takes at least 30 minutes to return to her normal self. Her blood sugar was 3.5 mmol/l during the most recent episode. Her parents have recently separated and she has a cousin who was treated for epilepsy.

What is the most likely diagnosis? (5 marks)

Select one answer only

A Benign seizures of childhood

B Complex partial seizures

C Fabricated seizure

D Hypoglycaemia

E Juvenile myoclonic epilepsy

F Long QT syndrome



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A 6 week old breast-fed baby girl presents with vomiting, poor feeding and lethargy of 1 day duration. She was born at term weighing 2.95 kg and was investigated for neonatal convulsions associated with hypoglycaemia that settled after the first week. She had been discharged home aged 10 days.

On initial examination, she was noted to have a capillary refill time of 4 seconds, temperature of 39°C, was jaundiced and her spleen was just palpable. Blood cultures grew E.coli and she responded well to intravenous antibiotics, but the jaundice persisted.

Current investigation shows a total bilirubin of 159 µmol/l (conjugated 56 µmol/l).

Which of the following additional investigations is most likely to help establish the diagnosis? (5 marks)

Select one answer only

A Brain MRI

B Galactose-1-phosphate uridyl transferase level

C Hepatitis serology

D HIDA liver scan

E Thyroid function tests

F Ultrasound of abdomen



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Study: Incidence of bacteraemia in infants and children with fever and petechiae.

Methods: Consecutive patients with a temperature of 38°C or higher and petechiae seen in the emergency department were prospectively enrolled. Our measures included (1) laboratory tests (leukocyte count, coagulation profile, blood culture, and cerebrospinal fluid bacterial culture); (2) a questionnaire requesting clinical data including general appearance, number and location of petechiae, and presence or absence of purpura; and (3) a follow-up telephone survey documenting health status.

Results: A total of 411 patients were enrolled, with 57.7% between 3 and 36 months of age. Eight patients (1.9%) had bacteraemia or clinical sepsis. None of the 357 well-appearing patients (95% confidence interval: 0.0% - 1.0%) had serious invasive bacteraemia. 53 patients appeared ill, including all 6 with serious invasive bacteraemia. An ill appearance of the child had a sensitivity of 1.00 (95% confidence interval: 0.60 - 1.00) and a leukocyte count of 15,000 or greater, or of less than 5000, had a sensitivity of 1.0 (95% confidence interval: 0.53 - 1.00) for detecting serious invasive bacteraemia. All children with bacteraemia had purpura.

Adapted from: Mandl KD, Stack AM, Fleisher GR. *J Pediatr* 1997 Sep;131(3):398-404

Which one of the following statements is the most reasonable conclusion to be drawn from this study? (5 marks)

Select one answer only

A All febrile children with petechiae should have a white blood cell count performed.

B Assessment of whether the child looks ill is a highly sensitive test for serious invasive bacteraemia.

C Assessment of whether the child looks ill is a highly specific test.

D Children with a white cell count between 5,000 and 15,000 should be allowed home.

E The white cell count is an accurate test for bacteraemia.

F These results do not apply to the UK because the study was performed in the United States.



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You will be presented with three clinical scenarios and a list of endocrine diagnoses.

Type in one answer only for each of the 3 clinical scenarios below

Each question is worth 3 marks

Note: each answer may be used more than once.

(EMQ 0005 SP)

A 2 year old girl with isolated bilateral breast development.

Select the most likely diagnosis from the list below: (3 marks)

A Constitutional growth and pubertal delay

B Growth hormone deficiency

C Hypothalamic hamartoma

D Idiopathic precocious puberty

E Klinefelter syndrome

F McCune – Albright syndrome

G Premature adrenarche

H Premature thelarche

I Testicular tumour

J Turner syndrome

Please add the correct answer in the box then continue to scroll down to the next part of the question

(EMQ 0005 SP a)

A 14 year old girl with short stature, delayed menarche and a cardiac murmur.

Select the most likely diagnosis from the list below: (3 marks)

A Constitutional growth and pubertal delay

B Growth hormone deficiency

C Hypothalamic hamartoma



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D Idiopathic precocious puberty

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E Klinefelter syndrome

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F McCune – Albright syndrome

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G Premature adrenarche

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H Premature thelarche

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I Testicular tumour

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J Turner syndrome

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Please add the correct answer in the box then continue to scroll down to the next part of the question

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(EMQ 0005 SP b)

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A 14 year old boy with short stature and a family history of delayed puberty.

Select the most likely diagnosis from the list below: (3 marks)

A Constitutional growth and pubertal delay

B Growth hormone deficiency

C Hypothalamic hamartoma

D Idiopathic precocious puberty

E Klinefelter syndrome

F McCune – Albright syndrome

G Premature adrenarche

H Premature thelarche

I Testicular tumour

J Turner syndrome

Please add the correct answer in the box

(EMQ 0005 SP c)



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A 6 year old boy was born with truncus arteriosus and had surgery when 3 days old. He made good post-operative recovery and continued with annual follow-up. He presents with isolated lesions on his toes (as shown in the image below) that had developed over the last 24 hours.

On examination, he has a temperature of 37.8°C, is centrally pink and there is no obvious changes to the known cardiac findings.

What is the most likely diagnosis? (4 marks)

Select one answer only

Clicking on the image below will open it in a new window, allowing it to be moved around the screen



A Deep venous thrombosis

B Idiopathic thrombocytopenic purpura

C Meningococcal septicaemia

D Protein S deficiency

E Subacute bacterial endocarditis

F Systemic lupus erythematosus

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A 4 year old boy presents with a 2 month history of progressive lethargy and pallor. He had previously been well and very active. Over the last 2 days he had become distressed when moved or cuddled by his parents. His appetite had reduced but he still managed to drink well and pass normal amounts of urine. He had an episodic temperature, which at times reached 38.5°C, over the 2 days prior to admission.

Examination shows him to be pale, withdrawn and unwilling to move his arms and legs. He has palpable lymph nodes in the cervical chain, left supraclavicular area, axillae and both inguinal areas. He is tachycardic and has a 2/6 soft ejection systolic murmur at the lower left sternal edge. Examination of the abdomen identifies a mass in the right upper quadrant measuring 10 x 7 cm.

What investigation will lead to a definitive diagnosis of the above presentation? (4 marks)

Select one answer only

A Blood cultures

B Cardiac echo

C Full blood count

D Liver function tests

E Serum alpha fetoprotein

F Urinary catecholamines



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A 7 year old girl was referred by GP with history of recurrent abdominal pain for 4 months. She has previously been treated for constipation with laxatives. She is able to pass soft stools without medication but her parents have noted fresh blood with stools during the last 2 weeks. Her mother is concerned that she appears to have lost weight recently and is less interested in sports.

Investigations:

haemoglobin	92 g/l	(115 - 140)
MCV	65 fl	(77 - 91)
CRP	83 mg/l	(< 5 mg/l)
albumin	32 g/l	(35 - 50)
faecal calprotectin	175 µg	

What is the most likely diagnosis? (4 marks)

Select one answer only

(7009 SP)

- A
- B
- C
- D
- E

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This question consists of two parts. Please answer both parts of the questions.

An 8 month old male infant was referred because of non-bilious vomiting. His GP had seen him frequently for constipation over the last few months.

Examination reveals a thin, non-dysmorphic infant weighing 6.8 kg (0.4th centile). He has a scaphoid abdomen and his capillary refill time is 3 seconds. General examination is otherwise unremarkable. His test results are shown below.

What is the most likely diagnosis? (6 marks)

Select one answer only

Clicking on the data chart below will open it in a new window, allowing it to be moved around the screen

Please continue to scroll down to the next part of the question

(D 0011 SP a)

Investigations:

Blood

haemoglobin	122 g/l	(110 - 140)
white cell count	$13 \times 10^9/l$	(6.0 - 15.0)
neutrophils	$9.4 \times 10^9/l$	(1.5 - 8.0)
lymphocytes	$3.6 \times 10^9/l$	(4.0 - 10.0)
platelets	$373 \times 10^9/l$	(150 - 450)
sodium	154 mmol/l	(133 - 146)
potassium	3.8 mmol/l	(3.5 - 5.5)
urea	6.0 mmol/l	(0.8 - 5.5)

Urine

microscopy	no red cells no white cells no casts
osmolality	180 mOsm/kg

A Chronic renal failure

B Diabetes insipidus

C Gastro-oesophageal reflux

D Hirschprung disease

E Renal tubular acidosis

F Type 1 diabetes

G Urinary tract infection

Which of the following is the most appropriate test to confirm the diagnosis? (5 marks)

Select one answer only

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microscopy

no red cells
no white cells
no casts
180 mOsm/kg

osmolality

A Chronic renal failure

B Diabetes insipidus

C Gastro-oesophageal reflux

D Hirschprung disease

E Renal tubular acidosis

F Type 1 diabetes

G Urinary tract infection

Which of the following is the most appropriate test to confirm the diagnosis? (5 marks)

Select one answer only

(D 0011 SP b)

A Blood sugar

B Oesophageal pH study

C Plasma aldosterone level

D Plasma osmolality

E Rectal biopsy

F Response to DDAVP

G Urinary pH



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A 10 year old Somali girl developed an abscess in the wound of a compound fracture from which E.coli was cultured. Her test results are shown below.

What is the most likely cause of the anaemia? (6 marks)

Select one answer only

Clicking on the data chart below will open it in a new window, allowing it to be moved around the screen

Investigations:

Blood

haemoglobin	68 g/l	(115 - 140)
MCV	82.3 fl	(77 - 91)
PCV	0.22	(0.35 - 0.48)
MCHC	30.6 g/dl	(32 - 35)
white cell count	12.3 x 10 ⁹ /l	(5.0 - 12.0)
neutrophils	9.1 x 10 ⁹ /l	(2.0 - 6.0)
lymphocytes	2.3 x 10 ⁹ /l	(1.5 - 7.0)
monocytes	0.9 x 10 ⁹ /l	(0.2 - 1.2)
reticulocytes	5.5%	
platelets	79 x 10 ⁹ /l	(150 - 450)
blood film	anisocytosis with microcytes and burr cells	
Coombs test	negative	

Urine microscopy	200 red blood cells x 10 ⁶ /l
	granular casts

- A
- B
- C
- D
- E
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A 13 year old girl presents with poor diabetic control and feeling unwell. She was admitted with diabetic ketoacidosis. Initial blood glucose is 27 mmol/l with a pH of 7.0. She responded well to fluids, resuscitation and insulin and her blood glucose falls to 10 mmol/l.

Twelve hours after admission, her level of consciousness deteriorated and her pulse dropped to 70/minute.

What is the most likely diagnosis? (4 marks)

Select one answer only

(7005 SP)

A Addison's disease

B Aspirin toxicity

C Cerebral oedema

D Diabetic ketoacidosis

E Reye syndrome



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This question consists of two parts. Please answer both parts of the questions.

This is a MAG 3 renogram of a boy with recurrent urinary tract infection.

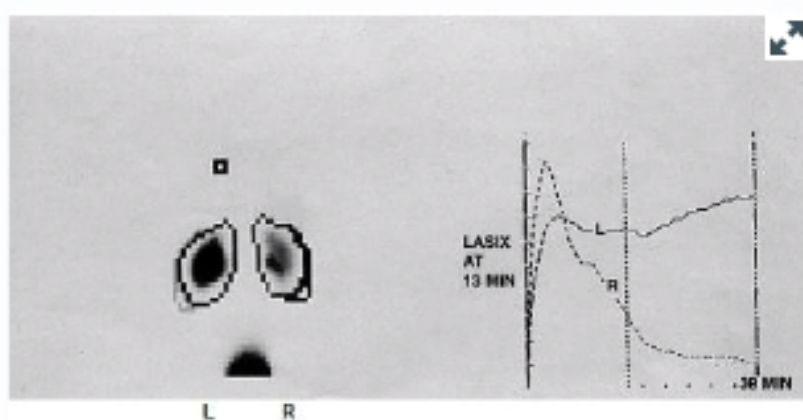
What important structural abnormality is shown? (5 marks)

Select one answer only

Clicking on the renogram below will open it in a new window, allowing it to be moved around the screen

Please continue to scroll down to the next part of the question

(X 0015 SP a)



A Bilateral pelvi-ureteric junction obstruction

B Bilateral renal cortical scarring

C Dilated left renal pelvis

D Dilated right renal pelvis

E Left renal cortical scarring

F Right pelvi-ureteric obstruction

G Right renal cortical scarring

H Posterior urethral valves

What functional abnormality is shown? (5 marks)

Select one answer only

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A Bilateral pelvi-ureteric junction obstruction

B Bilateral renal cortical scarring

C Dilated left renal pelvis

D Dilated right renal pelvis

E Left renal cortical scarring

F Right pelvi-ureteric obstruction

G Right renal cortical scarring

H Posterior urethral valves

What functional abnormality is shown? (5 marks)

Select one answer only

(X 0015 SP b)

A Bilaterally delayed excretion

B Bilateral ureteric reflux

C Delayed excretion on left despite furosemide

D Delayed excretion on right despite furosemide

E Left ureteric reflux

F Poor uptake on the left

G Poor uptake on the right

H Right ureteric reflux



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This 3 year old Romanian child, who has recently arrived in the UK, was noted to have an abnormality of his nail bed.

In which of the following is this physical sign a clinical feature? (4 marks)

Select one answer only

Clicking on the image below will open it in a new window, allowing it to be moved around the screen

(C 2129 SP)



A Congenital cystic adenomatoid malformation

B Eisenmenger syndrome

C Homozygous sickle cell disease

D Polycystic kidneys

E Pulmonary stenosis

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A 15 year old girl with a BMI of 15 (0.4 centile) had been diagnosed with ileocaecal Crohn's disease. She was to be commenced on enteral nutrition given by nasogastric tube in order to achieve remission. Her test results are shown below.

Which of the following is the most important action to take before commencing the enteral feeding regime? (4 marks)

Select one answer only

Clicking on the data chart below will open it in a new window, allowing it to be moved around the screen

Investigations:

Blood

haemoglobin	93 g/l	(115 - 165)
white cell count	11.3 x 10 ⁹ /l	(3.0 - 10.0)
normal differential		
platelets	275 x 10 ⁹ /l	(150 - 400)
ferritin	15 µg/l	(12 - 200)
sodium	133 mmol/l	(135 - 146)
choride	95 mmol/l	(95 - 106)
potassium	2.9 mmol/l	(3.5 - 5.3)
urea	6.0 mmol/l	(2.5 - 7.8)
creatinine	55 µmol/l	(60 - 120)
phosphate	1.3 mmol/l	(0.9 - 1.8)

A Commence oral iron therapy

B Correct hypokalaemia

C Give intramuscular vitamin B12

D Start oral multivitamin therapy

E Transfuse haemoglobin to 120 g/l

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Select the two most likely causes for the appearance of this 4 year old boy. (6 marks)

Select two answers only

Clicking on the image below will open it in a new window, allowing it to be moved around the screen

(C 0022 SP)



A Acute nephritis

B Bilateral periorbital cellulitis

C Cardiac failure

D Henoch-Schönlein purpura

E Hereditary angio-oedema

F Myotonia

G Nephrotic syndrome

H Non-accidental injury

I Superior vena cava obstruction

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An 8 year old girl was diagnosed as having cystic fibrosis on neonatal screening. She had been treated with oral flucloxacillin for intermittent *Staphylococcus aureus* grown from cough swabs in the clinic. The most recent sample has grown *Pseudomonas aeruginosa*.

Which of the following is the most appropriate management? (4 marks)

Select one answer only

(SH 0030 SP)

A Inhaled tobramycin for 3 weeks

B IV ceftazidime for 14 days

C Nebulised Dnase for 3 weeks

D Nebulised hypertonic saline for 3 weeks

E Oral ciprofloxacin and nebulised colomycin for 3 months



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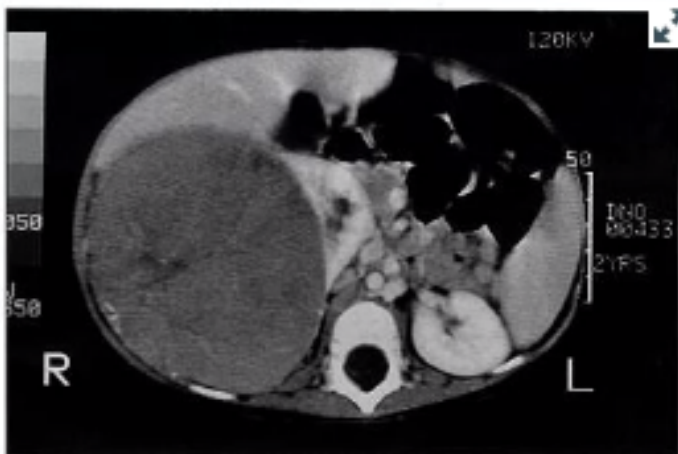
A 4 year old child presented with an abdominal mass.

What two abnormalities are seen on the CT scan below with IV contrast? (6 marks)

Select two answers only

Clicking on the CT scan below will open it in a new window, allowing it to be moved around the screen

(X 0014 SP)



A Displaced right kidney

B Megacolon

C Multi-cystic right kidney

D Solid tumour of the liver

E Solid tumour of the right adrenal gland

F Solid tumour of the right kidney

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You will be presented with three clinical scenarios with a list of investigations for infants with scrotal problems.

Type in one answer only for each of the 3 clinical scenarios below

Each question is worth 3 marks

Note: each answer may be used more than once

(EMQ 0019 SP)

At 6 week check, a baby was noted to have an empty scrotum. A mobile non-tender mass is palpable high in the inguinal canal.

Which should be the next step in the immediate management? (3 marks)

- A 17-hydroxyprogesterone assay
- B 21-alpha-hydroxylase assay
- C CT abdomen
- D DNA analysis for CYP21 gene mutation
- E GP review at 6 months
- F Karyotyping
- G Referral to clinical geneticist
- H Referral to paediatric surgeon
- I Ultrasound of abdomen
- J Urea and electrolytes

Please add the correct answer in the box below then continue to scroll down to the next part of the question

(EMQ 0019 SP a)

At 6 week check a baby was noted to have an empty scrotum and hypospadias.

Which should be the next step in the immediate management? (3 marks)

- A 17-hydroxyprogesterone assay
- B 21-alpha-hydroxylase assay



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C CT abdomen

D DNA analysis for CYP21 gene mutation

E GP review at 6 months

F Karyotyping

G Referral to clinical geneticist

H Referral to paediatric surgeon

I Ultrasound of abdomen

J Urea and electrolytes

Please add the correct answer in the box below then continue to scroll down to the next part of the question

(EMQ 0019 SP b)

At 6 week check a baby was noted to have a firm tender swelling in the right inguinal canal. Both testes are in the scrotum.

Which should be the next step in the immediate management? (3 marks)

A 17-hydroxyprogesterone assay

B 21-alpha hydroxylase assay

C CT abdomen

D DNA analysis for CYP21 gene mutation

E GP review at 6 months

F Karyotyping

G Referral to clinical geneticist

H Referral to paediatric surgeon

I Ultrasound of abdomen

J Urea and electrolytes

Please add the correct answer in the box below

(EMQ 0019 SP c)



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A 2 year old boy became coryzal and lethargic. He refused food saying his throat was sore and his fluid intake was reduced. He continued to pass urine although his mother states that it was darker in colour. He subsequently became drowsy and then had a generalised tonic-clonic seizure lasting about 4 minutes. He remained drowsy after this and was brought to hospital.

On observation in the emergency department, it is noted that he is rousable and aware of his mother's presence. His temperature is 38.2°C, heart rate 100 and BP is 90/60. Blood tests are undertaken and the results are shown below.

Investigations:

glucose	4.6 mmol/l	(3.0 - 6.0)
sodium	137 mmol/l	(133 - 146)
potassium	4.5 mmol/l	(3.5 - 5.5)
creatinine	30 mmol/l	(13 - 39)
urea	2.5 mmol/l	(2.5 - 6.5)

What is the most likely diagnosis? (4 marks)

Select one answer only

A Addison disease

B Benign focal epilepsy of childhood

C Febrile convulsion

D Glycogen storage disease

E Hypopituitarism

F Medium chain acyl-CoA dehydrogenase deficiency (MCAD)



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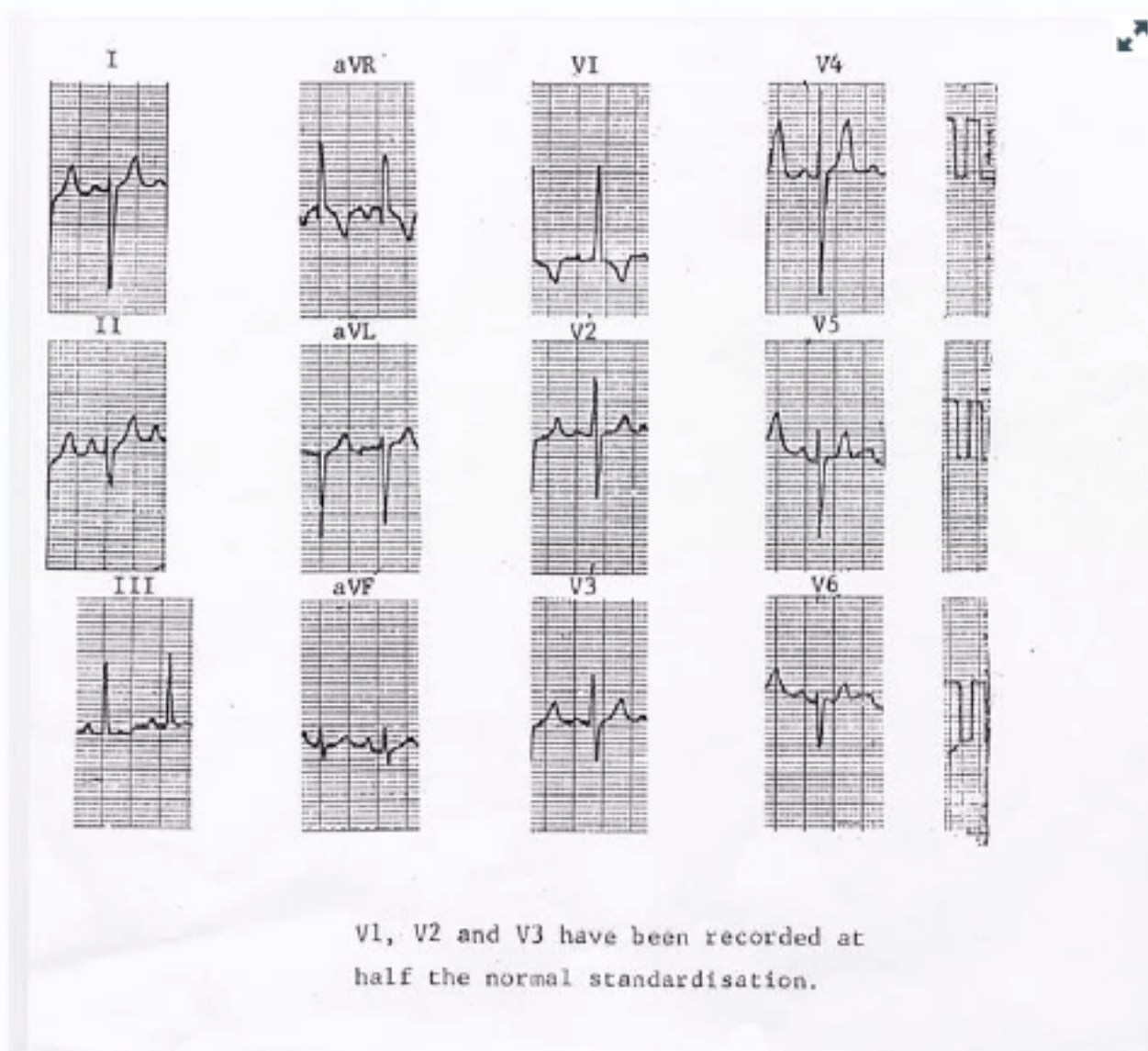
This ECG is from an active acyanotic 5 year old child who has signs of heart disease.

What is the most likely diagnosis? (5 marks)

Select one answer only

Clicking on the ECG below will open it in a new window, allowing it to be moved around the screen

(DE 252 SP)



A Atrial septal defect

B Fallot's tetralogy

C Patent arterial duct (ductus arteriosus)

D Pulmonary stenosis

E Ventricular septal defect



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A 14 year old boy with type 1 diabetes presents having had a series of collapses at school. His teacher provides a written history of the boy "going down like a sack of potatoes". The boy has no recall of events during these collapses.

His neurological, cardiac examination and blood pressure are all normal.

What is the most likely diagnosis? (5 marks)

Select one answer only

(7008 SP)

A Aortic stenosis

B Benign seizures of childhood

C Complex partial seizures

D Hypoglycaemia

E Juvenile myoclonic epilepsy

F Long QT syndrome



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A 5 year old boy suffering from painless daily rectal bleeding for the past 6 months was referred by his GP. The episodes occurred at any time of day and it was estimated that there was 1-2 teaspoons of fresh blood and visible mucous with each stool. There is no history of abdominal pain, weight loss, lethargy or loss of appetite. There is no relevant family history.

well child with anemia due to the bleeding from the polyp

Investigations:

haemoglobin	91 g/l	(115 - 140)
MCV	63 fl	(77 - 91)
CRP	7 mg/l	(<5)
albumin	39 g/l	(35 - 50)

Which of the following is the most likely explanation for his symptoms? (4 marks)

Select one answer only

(7007 SP)

- A Anal fissure
- B Campylobacter enteritis
- C Crohn's colitis
- D Juvenile colonic polyp
- E Meckel's diverticulum

Applied Knowledge in Practice – Sample Paper 2

Answer Key

Question 1

Question No: (C 0012 SP)

Answer: *E - Offer reassurance*

Reasoning:

The appearances are sometimes described as a 'geographic tongue' – where there are areas of sharply demarcated, irregular red patches with surrounding white plaques. These features are restricted to the dorsum of the tongue. The aetiology remains unclear.

Answer A is wrong

The papillae on the tongue are visible and not distorted and there is no evidence of erythema that would suggest malignancy or infection. Biopsy is not indicated.

Answer B is wrong

From the history, the child is well and therefore HIV would not be likely.

Answer C is wrong

This question wants you to be confident and reassure – a follow up review indicates you are unsure!!

Answer D is wrong

Nystatin would be indicated if this was a fungal infection but again the papillae would be covered and usually includes the buccal mucosa (though not visible here).

Answer E is the best answer

Also known as Benign Migratory Glossitis. The white coating is desquamating material. You can reassure.

Tip: research images for 'Strawberry tongue', oral candida, lichen planus.

Further Reading

Nelson Textbook of Paediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXXI. The Skin

Question 2

Question No: (D 0009 SP)

Answer: *B - Low maternal platelet count in pregnancy*

Reasoning:

This child presents with bruising on day 3 of life.

Answer A is wrong

A positive family history is indicative of an inherited clotting problem and not a thrombocytopaenia.

Answer B is the best answer

In maternal ITP, Neonatal Alloimmune Thrombocytopenia can occur due to trans-placental transfer of maternal antiplatelet autoantibodies.

Answer C is wrong

The given history does not indicate that mother was taking anti-epileptic medication

Answer D is wrong

The child is well so sepsis is unlikely to be the cause of the low platelets.

Answer E is wrong

Vitamin K is given to protect against haemorrhagic disease of the newborn – a problem of clotting, not thrombocytopaenia.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXI. Diseases of the Blood

Question 3

Question No: (EBM 0011 SP)

Answer: *D - Short course treatment resulted in a significantly reduced length of hospital stay.*

Answer: *F - There was no difference in the level of neurological complications between treatment groups.*

Reasoning:

'Odds ratios' compare the odds of a particular outcome occurring following one intervention and the outcome occurring following another intervention [OR=Outcome with Intervention 1/ Outcome with Intervention 2]. If the ratio equals 1 (or the confidence interval given spans across 1) then there is no difference in the two interventions.

Answer A is wrong

The OR confidence interval crosses 1 and therefore there is no observed difference between the two interventions.

Answer B is wrong

If the paper has already been published in a recognised journal, it is likely that the methodology would have been reviewed and therefore any answer that indicates a poor design of the trial is unlikely to be correct. This is a general statement and not based on the data provided so should be excluded

Answer C is wrong

There is no Further Reading to this point in the provided data and so not possible to comment.

Answer D is the best answer

The difference in length of stay is -2.17 days and the CI values are all negative indicating a reduced duration of stay – and, intuitively you would suggest this is true!!

Answer E is wrong

You are not provided with details of the clinical outcome just information of the possible long-term effects. You cannot propose a change of practice from the details given.

Answer F is the best answer

The provided OR crosses 1 and that suggests no difference.

Question 4

Question No: (7003 SP)

Answers: *A - Benign seizures of childhood*

Reasoning:

Answer A is the best answer

The seizures are often at night and are infrequent in nature. The prognosis is very good and seizures will resolve in most children often by the early teenage years.

Answer B is wrong

The history does not describe seizures with abnormal, coordinated actions

Answer C is wrong

The history given does not support this diagnosis

Answer D is wrong

The blood glucose was within the normal range when tested during a recent episode

Answer E is wrong

The episodes are described as 'shaking' and not 'jerking'

Answer F is wrong

The patient would usually present with sudden collapse.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016.
Chapter XXVII. The Nervous System

Question 5

Question No: (D 0010 SP)

Answer: *B - Galactose-1-phosphate uridyl transferase level*

Reasoning:

A 6 week old baby girl presenting with poor feeding, vomiting, hypoglycaemia, seizures and a conjugated hyperbilirubinaemia. Raise the likelihood of a metabolic condition.

Answer A is wrong

Intracranial abnormalities would not give a conjugated hyperbilirubinaemia.

Answer B is the best answer

This history would explain both the early neonatal events and the recent presentation. The symptoms are all consistent with a diagnosis of galactosaemia.

Answer C is wrong

Although hepatitis may give some of the findings listed, it would not explain the early neonatal history.

Answer D is wrong

A biliary atresia needs to be considered but such a diagnosis would not explain the early neonatal history.

Answer E is wrong

Congenital hypothyroidism produces an unconjugated hyperbilirubinaemia

Answer F is wrong

Would not explain the early neonatal problems and will give limited information on anatomy.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XI. Metabolic Disorders

Question 6

Question No: (EBM 0043 SP)

Answers: *B - Assessment of whether the child looks ill is a highly sensitive test for serious invasive bacteraemia.*

Reasoning:

Sensitivity could also be called the 'True Positive Rate' – the ability of the test to identify those with the condition of interest. In this example the condition of interest is 'bacteraemia'.

Specificity could also be called the 'True Negative Rate' – the ability of the test to exclude those who do not have the condition of interest.

Answer A is wrong

The white count is a valuable assessment tool but that is not the same as – 'All febrile children with petechiae should have a white blood cell count performed'.

Answer B is the best answer

'53 patients appeared ill, including all 6 with serious invasive bacteraemia'. Observation was able to identify all with bacteraemia though the false positive rate was high.

Answer C is wrong

The defined outcome is 'bacteraemia' and the question asks about 'specificity' of 'looking ill'. So, could the 'test' (clinical assessment) exclude those with the condition (bacteraemia). Eight children had bacteraemia but 53 were judged as unwell. The test is not specific.

Answer D is wrong

Although the study suggests that those children with white cell count between 5,000 and 15,000 are not likely to have a bacteraemia it does not lead onto saying they should be allowed home. Eight children had bacteraemia but 53 looked unwell.

Answer E is wrong

Low or high white counts were sensitive of bacteraemia but there is no comment about the relationship of intermediate white count. Therefore, one cannot make any decision on management based on this unreported result.

Answer F is wrong

It is always important to ask whether the population studied is the same as the population encountered in one's clinical practice. In this case children in the UK are very similar to those in the United States.

Question 7 a

Question No: (EMQ 0005 SP a)

Answer: *H - Premature thelarche (common, self-limiting)*

Reasoning:

Answer A is wrong

Constitutional delay of growth and puberty causes delay of puberty.

Answer B is wrong

Growth hormone deficiency causes delay of puberty.

Answer C is wrong

Hypothalamic hamartoma is associated with abnormal, early puberty with both breast and pubic hair development.

Answer D is wrong

Idiopathic precocious puberty is associated with abnormal, early puberty with both breast and pubic hair development.

Answer E is wrong

Klinefelter syndrome will only affect boys

Answer F is wrong

McCune-Albright Syndrome is associated with abnormal, early puberty with both breast and pubic hair development.

Answer G is wrong

Premature adrenarche is due to mild androgen production from the zona reticulosa of the adrenal gland as it matures and so causes pubic hair that is almost always confined to the vulva.

Answer H is the best answer

Answer I is wrong

Testicular tumour will only affect boys

Answer J is wrong

Turner Syndrome causes delayed puberty in girls.

Further Reading

UpToDate <https://www.uptodate.com/contents/definition-etiology-and-evaluation-of-precocious-puberty> Authors: Harrington J; Palmert MR. [Accessed May 2018]

Question 7 b

Question No: (EMQ 0005 SP b)

Answer: *J - Turner syndrome*

Reasoning:

By definition, menarche is not considered to be delayed until the age of 16 years. It comes towards the end of normal puberty ie 2-3 years following the first signs of secondary sexual characteristics.

Answer A is wrong

Constitutional delay is possible especially if the parents are short but this information is not provided in the question stem.

Answer B is wrong

Growth hormone deficiency is possible if a positive family history was provided.

Answer C is wrong

Hypothalamic hamartoma is associated with abnormal, early puberty with both breast and pubic hair development.

Answer D is wrong

Idiopathic precocious puberty is associated with abnormal, early puberty.

Answer E is wrong

Klinefelter syndrome will only affect boys

Answer F is wrong

McCune-Albright Syndrome is associated with abnormal, early puberty.

Answer G is wrong

Premature adrenarche is due to mild androgen production from the zona reticulosa of the adrenal gland as it matures and so causes pubic hair that is almost always confined to the vulva.

Answer H is wrong

Breast development is not mentioned in the history.

Answer I is wrong

Testicular tumour will only affect boys

Answer J is the best answer

Further Reading

UpToDate: <https://www.uptodate.com/contents/clinical-manifestations-and-diagnosis-of-turner-syndrome> Author: Backeljauw P

UpToDate: <https://www.uptodate.com/contents/approach-to-the-patient-with-delayed-puberty> Authors: Crowley WF; Pitteloud N. [Accessed May 2018]

Question 7 c

Question No: (EMQ 0005 SP c

Answer: *A - Constitutional growth and pubertal delay*

Reasoning:

Of the causes of delayed puberty in boys on this list the possible answers are Klinefelter syndrome, growth hormone (GH) deficiency and constitutional delay of growth and puberty (CDGP).

Answer A is the best answer

The question asks for the 'most likely diagnosis' which is therefore the common CDGP. This can be a source of misery for affected young men and responds well to a short course of low dose testosterone.

Answer B is wrong

GH deficiency is possible but very rare to be diagnosed at this age.

Answer C is wrong

Hypothalamic hamartoma is associated with abnormal, early puberty.

Answer D is wrong

Associated with early puberty.

Answer E is wrong

Klinefelter characteristically causes disproportionate relative tall stature.

Answer F is wrong

Associated with early puberty.

Answer G is wrong

The history is about delay in puberty.

Answer H is wrong

The history is about delay in puberty.

Answer I is wrong

Testicular problem not mentioned in history

Answer J is wrong

Found only in females

Further Reading

UpToDate: <https://www.uptodate.com/contents/approach-to-the-patient-with-delayed-puberty> Authors: Crowley WF; Pitteloud N. [Accessed May 2018]

Question 8

Question No: (C 0017 SP)

Answer: *E - Subacute bacterial endocarditis*

Reasoning:

Dark purple/black (ischaemic) lesions on the big toe and on the plantar surface of the foot. These are appearances of the effect of vascular occlusion.

Answer A is wrong

Deep vein thrombosis would usually lead to swelling of the distal limb and this is not evident here

Answer B is wrong

The resulting lesions from ITP would be distributed throughout the torso and limbs

Answer C is wrong

Meningococcal septicaemia causing such lesions would be indicative of overwhelming and rapidly progressive infection. The child would be septic and significantly unwell.

Answer D is wrong

The individual with Protein S deficiency produces thrombophilia and an increased risk of thromboembolic events. Most events are DVT, pulmonary embolism or cerebral thrombosis.

Answer E is the best answer

This young boy is at risk of developing Subacute bacterial endocarditis following his surgical repair and these lesions would be consistent with embolic thrombi from the endocarditis. The past history makes this the more likely explanation.

Answer F is wrong

There is no history of the characteristic butterfly rash or joint pain to make this a possible diagnosis.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 9

Question No: (7004 SP)

Answer: *F - Urinary catecholamines*

Reasoning:

Answer A is wrong

Sepsis must always be considered and may explain many of the features here. An intra-abdominal abscess could produce a mass, one would expect the child to be toxic.

Answer B is wrong

An infective cardiac lesion would not produce an abdominal mass and the cardiac murmur fits the description of a flow murmur.

Answer C is wrong

The FBC is likely to be abnormal but would not lead to a definitive diagnosis of the underlying aetiology.

Answer D is wrong

The liver function tests are likely to be abnormal but would not lead to a definitive diagnosis of the underlying aetiology.

Answer E is wrong

A raised alpha fetoprotein would be indicative of a hepatoblastoma but the abdominal lesion described is more that of discrete lesion rather than an enlarged liver.

Answer F is the best answer

The history and examination are highly suggestive of neuroblastoma. The pallor and the presence of left suprasternal lymph nodes (Virchow nodes) are indicative of metastatic disease. Raised urinary catecholamines are indicative of neuroblastoma although they can be negative in 5% of children.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XXII Cancer and Benign Tumours

Question 10

Question No: (7009 SP)

Answer: *B - Crohn's colitis*

Reasoning:

Most diagnoses are made on the history. This girl is chronically unwell – she has pain, weight loss and, importantly, no longer feels well enough to participate in sport. The blood tests show a microcytic anaemia, likely to be due to iron deficiency, plus evidence of inflammation somewhere with the high CRP. The low albumin also suggests chronic illness, especially in the bowel. Finally, the faecal calprotectin is high which is a very useful indicator of bowel inflammation when neutrophils migrate to the bowel wall. Therefore, the likeliest diagnosis is Crohns colitis.

Further Reading

UpToDate: <https://www.uptodate.com/contents/clinical-presentation-and-diagnosis-of-inflammatory-bowel-disease-in-children> Authors: Highuchi L; Bousvaros A; [Accessed May 2018]

Question 11 a

Question No: (D 0011 SP a)

Answers: B - *Diabetes insipidus*

Reasoning:

This baby is vomiting, has faltering growth and deranged biochemistry.

Answer A is wrong

The urea is slightly high but not in the range for chronic renal failure.

Answer B is the best answer

The urine osmolality is low in the face of a high serum. This is indicative of diabetes insipidus.

Answer C is wrong

The abnormally high sodium excludes GORD.

Answer D is wrong

The biochemistry is normal in Hirschprung and the abdomen is likely to be distended, not scaphoid.

Answer E is wrong

In pseudohypoaldosteronism, whatever the type, there is increased renal tubular sodium loss resulting in hyponatraemia and hyperkalaemia.

Further Reading

<https://www.uptodate.com/contents/hyponatremia-in-children>

Question 11 b

Question No: (D 0011 SP b)

Answers: F - *Response to DDAVP*

Reasoning:

Answer A is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer B is wrong.

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer C is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer D is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer E is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Answer F is the best answer

To make a diagnosis of diabetes insipidus (DI) requires a water deprivation test with, if there is evidence of on-going polyuria and weight loss, the assessment of the effect of a dose of desmopressin will help distinguish between central and nephrogenic DI. This test is dangerous and must only be undertaken by trained and experienced teams.

Answer G is wrong

Diagnosis of diabetes insipidus is confirmed by the water deprivation test.

Further Reading

UpToDate: <https://www.uptodate.com/contents/hyponatremia-in-children> Authors:
Somers MJ; Traum AZ. [Accessed May 2018]

Question 12

Question No: (D 1136 SP)

Answer: *E - Microangiopathic intravascular haemolysis*

Reasoning:

This is a normochromic, normocytic anaemia with thrombocytopenia and a slightly elevated white count. The blood film shows anisocytosis (red cells of different sizes) and burr cells (or echinocytes which show a crenelated membrane and are changes seen due to environmental causes).

Many of these answers can be excluded as the cause of the anaemia

Answer A is wrong

Acute lymphoblastic leukaemia (normal film, an appropriately elevated reticulocyte count indicating a responsive marrow and normal MCV –CHECK machine MCV of anisocytosis)

Answer B is wrong

Blood loss (nothing in the given history),

Answer C is wrong

HIV can give anaemia but you would expect the child to have a longer history of illness.

Answer D is wrong

Malaria would be identified on the film

Answer E is the best answer

Anaemia and thrombocytopaenia along with granular casts in urine indicate acute haemolysis - microangiopathic intravascular haemolysis.

Answer F is wrong

Sickle cell disease would be identified on the film.

Answer G is wrong

Thalassaemia would give a microcytic anaemia

Answer H is wrong

Tuberculosis would give a microcytic anaemia

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016.
Chapter XXI. Diseases of the Blood

Question 13

Qu No: (7005 SP)

Answer: *C - Cerebral oedema*

Reasoning:

The 13 year old girl admitted in DKA and is hyperglycaemic and acidotic. Progresses into coma after 12 hours of treatment as her blood glucose falls toward normal.

Answer A is wrong

Addison's disease is rare in children and young people but can lead to recurrent hypoglycaemia in patients with type 1 diabetes mellitus. There is insufficient information given to conclude that this diagnosis explains the change in clinical condition.

Answer B is wrong

Although acidotic there is nothing in history to suggest aspirin was administered. Aspirin is contraindicated in those under 16 years because of the increased risk of developing Reye's syndrome.

Answer C is the best answer

During resuscitation this child given fluids and there are rapid shifts of fluid between various body spaces during this time. Such fluid shifts can be rapid and lead to cerebral oedema, coning and death. Paediatric units would be expected to have clear and robust fluid replacement guidelines in place to ensure a slow and controlled rehydration.

Answer D is wrong

The glucose is returning to normal and one would not expect a clinical deterioration with a blood glucose of 10 mmol/l.

Answer E is wrong

Reye's syndrome - hepatic encephalopathy – would produce an encephalopathy often with confusion and seizures. No laboratory results are provided to suggest this diagnosis.

Further Reading

UpToDate: <https://www.uptodate.com/contents/cerebral-edema-in-children-with-diabetic-ketoacidosis> Author: Haymond MW; [Accessed March 2018]

Question 14 a

Question No: (X 0015 SP a)

Answers: C - Dilated left renal pelvis

Reasoning:

It is unlikely that you will be asked to interpret a MAG3 scan in clinical practice but an understanding of the investigation helps in explaining the results.

The image on the left of the picture shows the uptake of the radioactive tracer by renal tissue and represents the two kidneys and bladder when viewed from the back of the patient. The right kidney shows increased concentration in the pelvic area (dense black) whilst the left kidney has extensive area of dense tracer presence – a dilated pelvis.

The image on the right plots out the uptake and decay of the contrast as it passes through the two kidneys. There is rapid uptake by both kidneys following the injection of the contrast with the right kidney absorbing slightly more than the left. The contrast tracer is then cleared by the kidney into the bladder and at 13 minutes furosemide (Lasix) is injected to enhance that clearance. The left kidney fails to clear the tracer and the injection of furosemide (Lasix) leads to a clearance of water and a consequent concentration of the tracer within the kidney – shown as a slight elevation in the line of the graph. The abnormality shown is in the left kidney.

Answer A is wrong

There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer B is wrong

There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer C is the best answer

The left kidney has increased density of contrast material and the renal pelvis is dilated.

Answer D is wrong

The abnormality is in the left kidney – the right is normal.

Answer E is wrong

Scarring would be shown by patches of failed uptake of tracer by the parenchyma.

Answer F is wrong

The abnormality is in the left kidney – the right is normal.

Answer G is wrong

The abnormality is in the left kidney – the right is normal. Scarring would be shown by patches of failed uptake of tracer by the parenchyma.

Answer H is wrong

Posterior urethral valves would give a symmetrical appearance and show a delay in excretion in both kidneys in the graph on the right.

Question 14 b

Question No: (X 0015 SP b)

Answer: *C - Delayed excretion on left despite furosemide*

Reasoning:

Again any 'bilateral' answer is incorrect as is any answer including the right kidney. Uptake of tracer is good in both kidneys (indicated by the steep gradient of the graph). Ureteric reflux would show a delayed excretion of the tracer and a slower decay in the right hand graph.

Answer A is wrong

There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer B is wrong

There is an asymmetrical response so any 'bilateral' answer is incorrect

Answer C is the best answer

Answer D is wrong

The abnormality is in the left kidney – the right is normal.

Answer E is wrong

Ureteric reflux would show a delayed excretion of the tracer and a slower decay in the right hand graph – but decay would occur.

Answer F is wrong

The left kidney shows a prompt uptake of the tracer – it is the excretion which is abnormal

Answer G is wrong

The right kidney shows a prompt uptake of the tracer – it is the excretion which is abnormal

Answer H is wrong

The abnormality is in the left kidney – the right is normal.

Answer I is wrong

Ureteric reflux would show a delayed excretion of the tracer and a slower decay in the right hand graph.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016.
Chapter XXIII. Nephrology

Question 15

Question No: (C 2129 SP)

Answer: *B - Eisenmenger syndrome*

Reasoning:

The image shows digital clubbing ie loss of the angle between the nail-bed and the nail. It is a much more reliable sign of chronic hypoxia than cyanosis. Indeed, in this image it is difficult to ascertain whether the child is cyanosed! Furthermore, peripheral cyanosis is most commonly associated with reduced blood flow through the extremities, rather than central cyanosis.

Having decided that the child has finger clubbing, the next step is to look for a condition in the list of possible answers that causes central cyanosis. The main causes of clubbing in children are: Suppurative lung disease (cystic fibrosis, bronchiectasis), cyanotic heart disease, infective endocarditis, inflammatory bowel disease.

Answer A is wrong

Cystic adenomatoid malformation is usually unilateral. It may present in the neonatal period with respiratory distress. Smaller lesions may present later in childhood with recurrent respiratory infections.

Answer B is the best answer

Eisenmenger syndrome is caused by a ventricular septal defect leading to pulmonary hypertension and a right-to-left shunt causing central cyanosis and clubbing.

Answer C is wrong

Acute chest syndrome in a child with homozygous sickle disease may cause oxygen desaturation, but chronic hypoxia is not a feature, and children do not develop clubbing.

Answer D is wrong

Polycystic kidney disease may lead to chronic renal failure, but hypoxia and clubbing are not seen in this condition.

Answer E is wrong.

Pulmonary stenosis is a form of acyanotic congenital heart disease, and clubbing is not a feature.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 16

Question No: (D 2330 SP)

Answers: *B - Correct hypokalaemia*

Reasoning:

The patient has a low BMI for her age as a result of malabsorption from her Crohn's disease. The significant abnormalities in the investigations are anaemia and hypokalaemia although ferritin, sodium and chloride are all borderline low. The question asks which 'is the most important action to take before commencing the enteral feeding regime?' Hypokalaemia needs prompt correction – the others can wait. Furthermore, were she to start an enteral feeding regimen whilst malnourished child she will be at risk of developing 'Re-feeding syndrome'. One of the consequences would be a further fall in potassium levels – it is therefore crucial that hypokalemia is corrected before enteral feeding is started.

Answer A is wrong

Returning the haemoglobin with oral iron in a well individual will take months and 93 g/l is not life-threatening

Answer B is the best answer

Prompt correction is required.

Answer C is wrong

Although B12 deficiency is possible, it has not been demonstrated on the results provided.

Answer D is wrong

Not an urgent requirement

Answer E is wrong.

A haemoglobin of 93 g/l is not life-threatening

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter VIII. The Acutely Ill Child.

Question 17

Question No: (C 0022 SP)

Answer: *E - Hereditary angio-oedema*

Answer: *G - Nephrotic syndrome*

Reasoning:

The picture shows periorbital oedema without obvious inflammation or bruising. The most common causes of periorbital oedema in the UK are nephrotic syndrome and conditions presenting with allergy or anaphylaxis.

Answer A is wrong

Nephritis can lead to a nephrotic picture but proteinuria is uncommon (<1%).

Answer B is wrong

Cellulitis is unlikely as there is no obvious erythema in the periorbital area

Answer C is wrong

Cardiac failure can give systemic oedema but it is usually postural and one would need more clinical information about cardiac status.

Answer D is wrong

There is no history or clinical evidence of the characteristic rash.

Answer E the best answer

An acute anaphylactic reaction gives periorbital swelling.

Answer F is wrong

A spurious answer as myotonia may give a ptosis but this is oedema and not ptosis!

Answer G is the best answer

A nephrotic syndrome will lead to hypoalbuminaemia

Answer H is wrong

No bruising – another spurious answer

Answer I is wrong

SVC obstruction gives full facial swelling, cyanotic facial appearance and engorged veins.

Further Reading

Up to Date: <https://www.uptodate.com/contents/evaluation-and-management-of-edema-in-children> Valentini RP; [Accessed May 2018]

Question 18

Question No: (SH 0030 SP)

Answer: *E - Oral ciprofloxacin and nebulised colomycin for 3 months*

Reasoning:

The current practice in the management of Pseudomonas colonisation in patients with cystic fibrosis involves the use of 'duel therapy' – antibiotics given systemically and via a nebulised route.

Answer A is wrong

Single agent antibiotic only.

Answer B is wrong

Single agent antibiotic only.

Answer C is wrong

This option does not include an antibiotic.

Answer D is wrong

This option does not include an antibiotic.

Answer E is the best answer

Duel therapy with oral and nebulised antibiotics.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016.
Chapter XIX The Respiratory System

Question 19

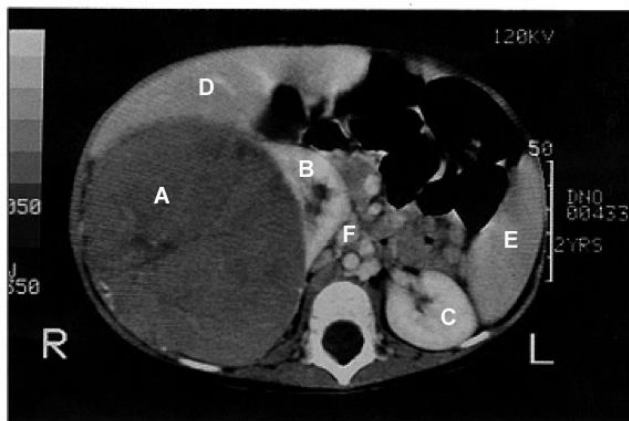
Question No: (X 0014 SP)

Answer: *A - Displaced right kidney*

Answer: *F - Solid tumour of the right kidney*

Reasoning:

IV contrast has been given which enhances vascular structures making them appear white. The view provided is a transverse section and the view is from the inferior position looking towards the head – the R and L markers indicate the appropriate sides.



The vertebra is evident posteriorly in the midline at the bottom of the image. Normal sized left kidney (C). Large mass (A) arising from the right kidney (B - enhanced). Liver (D) and spleen (E).

Answer A is the best answer

Answer B is wrong

The bowel gas pattern is of normal calibre although displaced slightly to the left.

Answer C is wrong

There are no cysts evident in the renal tissue (B or C).

Answer D is wrong

The area of liver visible is normal in appearance and is not displaced.

Answer E is wrong

An enlargement of the adrenal tissue (neuroblastoma) would displace the right kidney inferiorly but would maintain the normal renal shape.

Answer F is the best answer

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016.
Chapter XXII Cancer and Benign Tumours

Question 20a

Question No: (EMQ 0019 SP a)

Answer: *E - GP review at 6 months*

Reasoning:

This is not an uncommon scenario. Most testes that are undescended at birth and complete their descent within the first three to four months of life. Spontaneous descent is rare after six months of age. Clinically this is an otherwise normal baby boy. The likelihood is that the palpable mass is a testis and the other one is either retractile or intra-abdominal. Provided the rest of the genitalia are normal there is no urgency to do anything but wait and see what happens - and reassure the parents.

Further Reading

UpToDate: <https://www.uptodate.com/contents/undescended-testes-cryptorchidism-in-children-management> Authors: Cooper CS; Docimo SG; [Accessed May 2018]

Question 20b

Question No: (EMQ 0019 SP b)

Answer: *J - Urea and electrolytes*

Reasoning:

There are a number of reasons for this appearance, but the most dangerous is a virilised female baby with 21-hydroxylase deficiency. The urgent complications of this condition are hypoglycaemia and adrenal crisis, typically in the first two weeks of life. Thus, the NEXT step would be to check the electrolytes (and a blood sugar), before going on to perform more diagnostic tests.

Further Reading

UpToDate: <https://www.uptodate.com/contents/diagnosis-of-classic-congenital-adrenal-hyperplasia-due-to-21-hydroxylase-deficiency-in-infants-and-children> Author: Merke DP; [Accessed May 2018]

Question 20c

Question No: (EMQ 0019 SP c)

Answer: *H - Referral to paediatric surgeon*

Reasoning:

If the testes are in the scrotum, this is likely to be an inguinal hernia, and most of the other options are irrelevant. Since it is tender, it may be incarcerated or strangulated, and thus surgical referral is indicated.

Further Reading

UptoDate <https://www.uptodate.com/contents/inguinal-hernia-in-children> Author: Ramsook C; [Accessed May 2018]

Question 21

Question No: (7006 SP)

Answer: C - *Febrile convulsion*

Reasoning:

Answer A is wrong

Although adrenal insufficiency (Addison disease) can cause electrolyte disturbances which may lead to seizure activity, the results are not consistent with such a diagnosis (hypoglycaemia, hyponatraemia, hyperkalaemia).

Answer B is wrong

He has only had one episode so this is not any form of epilepsy. This episode does not fit the pattern of BFEC – recurrent, infrequent, often at night.

Answer C is the best answer

The child has an intercurrent illness and is mildly pyrexial when assessed though may have had a higher temperature around the time of the seizure. He is of the expected age for febrile seizure and has no abnormal neurological findings.

Answer D is wrong

There are no features in the history or blood results to support this diagnosis

Answer E is wrong

The clinical history is of an acute onset. The blood results are not consistent with this diagnosis.

Answer F is wrong

Children with MCAD do present during a period of fasting and poor oral intake and can have seizures. However, they have hypoglycaemia.

Further Reading

UpToDate: <https://www.uptodate.com/contents/seizures-and-epilepsy-in-children-classification-etiology-and-clinical-features> Author: Wilfong A. [Accessed March 2018]

Question 22

Question No: (DE 252 SP)

Answer: *D - Pulmonary stenosis*

Reasoning:

The ECG shows that the child is in sinus rhythm with a normal PR interval. There is marked right axis deviation of 180° as shown by an equiphase QRS complex in aVF with an upward QRS complex in aVR. There is also right ventricular hypertrophy with tall R waves over the right chest leads (note that V1, V2 and V3 have been recorded at half the normal standardisation).

Answer A is wrong

Ostium primum atrial septum defect is also a type of acyanotic congenital heart disease, but the ECG is typically associated with right bundle branch block and right axis deviation (without right ventricular hypertrophy) NB PRIMUM V SECUNDUM

Answer B is wrong.

In Fallot tetralogy the child would be cyanosed. The ECG shows right axis deviation and right ventricular hypertrophy. It would be extremely unlikely to see a 5 year old child with uncorrected tetralogy of Fallot in this country.

Answer C is wrong

In uncomplicated patent arterial duct, the child is usually pink and well, and there are no diagnostic ECG features.

Answer D is the best answer

Of the types of congenital heart disease listed, the only type of acyanotic heart disease associated with right axis deviation and right ventricular hypertrophy is pulmonary stenosis.

Answer E is wrong

A child with an uncomplicated ventricular septal defect is pink. The ECG is typically normal

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 23

Question No: (7008 SP)

Answer: *F - Long QT syndrome*

Reasoning:

Answer A is wrong

The cardiac examination was normal!

Answer B is wrong

The episodes do not occur at night

Answer C is wrong

The history does not describe seizures with abnormal, coordinated movements

Answer D is wrong

Hypoglycaemia may cause seizures and loss of consciousness, but the individual would experience symptoms such as sweating, anxiety, weakness and headaches before the loss of consciousness and is usually preceded by increasing drowsiness.

Answer E is wrong

The episodes are described as 'collapse' and not 'jerking'

Answer F is the best answer

Episodes of loss of consciousness during exertion suggest a cardiac disorder. Any child presenting in this way should have an ECG that will show a prolonged QTc of greater than 0.46 msec.

Further Reading

Nelson Textbook of Pediatrics. 20th Edition. Kliegman RM et al. Elsevier. Philadelphia 2016. Chapter XX. The Cardiovascular System

Question 24

Question No: (7007 SP)

Answer: D - *Juvenile colonic polyp*

Reasoning:

This boy is well and the bleeding is painless. He has a microcytic anaemia suggestive of iron deficiency so the bleeding is significant and over a prolonged period of time.

Answer A is wrong

Painless bleeding would make anal fissure very unlikely.

Answer B is wrong.

This is rather a prolonged duration of illness and the fresh blood indicates a bleeding point rather than widespread mucosal damage and inflammation.

Answer C is wrong

The normal albumin and CRP help exclude inflammatory bowel disease.

Answer D is the best answer

The presence of mucous and significant fresh bleeding makes the likeliest diagnosis a juvenile colonic polyp.

Answer E is wrong

This diagnosis is possible but most patients with complications from a Meckel's are younger and the blood rarely fresh

Further Reading

UpToDate: <https://www.uptodate.com/contents/lower-gastrointestinal-bleeding-in-children-causes-and-diagnostic-approach> Authors: Patel N; Kay M; [Accessed May 2018]

MRCPCH Applied Knowledge in Practice Paper 1 Introduction

Welcome to the MRCPCH Applied Knowledge in Practice Paper 1 examination for computer-based testing. If you are new to the MRCPCH computer-based examination, please note that some questions may appear in a slightly different format from how they did in paper format.

Please answer all of the parts of each of the questions. You will have 2 hours and 30 minutes to complete the examination (unless you have been granted additional time).

Some questions will have multiple parts to them, so please remember to scroll to the bottom of EACH question in order to view all parts. Please answer all parts of a question and check that you have entered an answer for all parts of each question.

You can adjust your display preferences at any point during the exam by clicking the preferences icon at the bottom of your screen.

You may find it helpful to eliminate some of the answer options to help you concentrate on others. You can do this by right-clicking over an answer option to strike a line through it. Right-clicking over it again will remove the line. *Please note that this does not apply to Extended Matching Questions.*

There is a highlight function that you may wish to use during the examination to help you to identify key text or questions to which you would like to return. You can do this by highlighting some text with your cursor and then clicking on the marker icon when it appears. The text will become highlighted in yellow and will remain highlighted throughout the remainder of the examination. *Please note that this highlight feature will not work with all questions.*

Extended Matching Questions (EMQ)

In this type of question, 3 scenarios are given, followed by a list of options. The option list might include a list of diagnoses, treatments, drugs or other management steps. For each scenario, you will be asked to choose the option from the list that is the most appropriate. Type the correct letter in the given box. Please note that the response entered is not case-sensitive, so it will not affect your result if the letter you type is in lower case or upper case.

Please Remember:

Please answer all parts of each question. If you do not answer all parts of a question, you will only be awarded marks for the parts of the question you have answered.

Please pay particular attention to this when answering the EMQ questions as they will have multiple parts.

It is strictly forbidden to talk to, read the work of, or attempt in any way to communicate with, other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work. The College has tools, which can identify copying of answers or collusion between candidates to share answers. If such a situation were to arise all identified parties will be investigated. Breaches of these instructions, or misbehaviour in any other way relating to this theory examination, including continuing to attempt to answer after the allotted time, may lead to suspension from the examination. Any attempt at copying or colluding to gain advantage may lead to permanent suspension from College examinations and notification to the GMC. If a candidate is suspected of malpractice during an examination they will be asked to leave the examination venue by the Chief Invigilator.

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Royal College of Paediatrics and Child Health
MRCPCH PART II EXAMINATION

PAPER 1

1. Complete the following:

Your full name (BLOCK LETTERS)

.....

RCPCH Number

Signature

.....

2. Please check that your surname (family name) is correct on the **Answer Sheet**.

Do not write anything on the Answer sheet other than your response to each question.

3. Answer all of the questions.

4. It is strictly forbidden to talk to, read the work of, or attempt in any way to communicate with, other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work. The College has tools which can identify copying of answers or collusion between candidates to share answers. In any situation the suspicion of guilt falls upon both parties until it can be proved otherwise. Breaches of these instructions, or misbehaviour in any other way, including continuing to write after the allotted time, may lead to suspension from the examination at the discretion of the invigilators. Serious breaches, such as cheating or colluding to gain advantage, could incur permanent suspension from College examinations.

5. Copyright law protects examination questions and the intellectual property of their authors. The unauthorised use of questions is a breach of copyright law.

6. Time allowed: 1 hour.

Questions 1 and 2

A 12-week-old baby was found by his parents blue and apparently lifeless in his cot after they had heard him choking. His father revived him with mouth-to-mouth resuscitation and brought him to the hospital.

He was born at 36 weeks gestation weighing 2.4 kg. He had no perinatal problems and fed well by bottle although he frequently regurgitated his feeds. His parents reported that he had been unwell with a cough for a few days before admission. His two siblings aged 5 years and 3 years had both been unwell recently with chest infections. Father had been unemployed for 18 months and there were considerable financial problems.

On examination he was pale and had a nasal discharge. His weight was on the 10th centile. Slight intercostal recession and a respiratory rate of 40/minute were noted together with scattered fine crepitations and a high pitched expiratory wheeze. The remainder of the examination was normal. Examination of the urine revealed no protein or sugar and no cells were seen on microscopy. A chest x-ray showed a degree of overinflation with no localised opacities.

1. What is the most useful investigation for establishing the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A echocardiogram
- B nasopharyngeal aspirate for respiratory viruses
- C pernasal swab
- D pH probe
- E sweat test

This question continues on the next page.

2. What is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A congenital cardiac abnormality
- B cystic fibrosis
- C gastro-oesophageal reflux
- D milk aspiration pneumonitis
- E pertussis infection
- F viral bronchiolitis

Question 3

This 9-month-old infant was cyanosed at birth and had a cardiac operation at 3 months of age.

3. What condition is shown here? (3 marks)

SELECT ONE ANSWER ONLY

- A left sided Horner's syndrome
- B left sided lower motor neurone facial palsy
- C left sided ptosis
- D left sided upper motor neurone facial palsy
- E right-sided Horner's syndrome
- F right-sided lower motor neurone facial palsy
- G right-sided ptosis
- H right-sided upper motor neurone facial palsy



Q3

Questions 4 and 5

An infant of 32 weeks gestation on his fourth day of life was well maintained on intermittent mandatory ventilation at 10 breaths per minute for respiratory distress syndrome. She was changed to nasal continuous positive airway pressure (CPAP) in 40% oxygen ($FiO_2 = 0.4$) at 5cm water pressure.

One hour after commencing nasal CPAP the following results were obtained from an arterial blood gas specimen:

pH	→ 7.20 (hydrogen ion concentration 62 nmol/l)
PaCO ₂	10.0 kPa (75mmHg)
PaO ₂	11.7 kPa (88mmHg)
Base excess	1.1 mmol/l
Standard bicarbonate	24 mmol/l
Actual bicarbonate	<u>28.8 mmol/l</u>

4. Select the best description for the acid-based abnormality. (2 marks)

SELECT ONE ANSWER ONLY

- A compensated metabolic acidosis
- B compensated respiratory acidosis
- C normal
- D partially compensated metabolic acidosis
- E partially compensated respiratory acidosis
- F uncompensated metabolic acidosis
- G uncompensated respiratory acidosis

This question continues on the next page.

5. What would be your next step in the management of this infant? (3 marks)

SELECT ONE ANSWER ONLY

- A chest X-ray
- B ensure nasal CPAP circuit is operating correctly
- C increase inspired oxygen concentration
- D reintubate and restart ventilation
- E transilluminate chest

Questions 6, 7 and 8

A 14-year-old boy of Bangladeshi origin was seen in the Accident & Emergency Department of a District General Hospital with a generalised convulsion. His parents said that he had complained for two weeks previously of mild headaches, which had occurred at different times of the day. At the age of 12 he was found to be sniffing glue but subsequently told his parents he had discontinued the practice. His progress at school was good and his behaviour had been normal.

On the afternoon of admission he had complained of a sudden generalised headache; despite this he had gone to see some friends but returned home with the headache. His mother had given him paracetamol. As he was sitting down to watch television, he became stiff and had a generalised convulsion.

The family called an ambulance and rectal diazepam was administered. He continued to fit and on arrival at the hospital, intravenous lorazepam was required to terminate the convulsion. He remained very drowsy and non-responsive.

On examination, there was some resistance to flexion of his neck but he was afebrile. His respirations were laboured, he was not cyanosed and was well perfused peripherally. His blood pressure was 160/90 mmHg. Examination of his heart, respiratory system and abdomen were normal.

His pupils were of equal size and both reacted sluggishly to light. Examination of the fundi showed no abnormalities; there was a generalised increase in tone in his limbs but no focal abnormal neurological signs.

6. What is the most appropriate investigation to establish the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A lumbar puncture
- B Mantoux test
- C CT scan
- D toxicology screen
- E urgent EEG

This question continues on the next page.

7. What are the two most appropriate forms of immediate management? (4 marks)

SELECT TWO ANSWERS ONLY

- A Administer intravenous antibiotics
- B Administer intravenous dexamethasone
- C Administer intravenous diuretic
- D Administer intravenous fluid bolus
- E Administer intravenous mannitol
- F Administer second-line anti-epileptic medication
- G Arrange for immediate intubation and ventilation
- H Discuss patient with a Paediatric Intensive Care Unit

8. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A cerebral tumour
- B drug overdose
- C epilepsy
- D meningitis
- E subarachnoid haemorrhage

Questions 9, 10, 11, 12, 13, 14 and 15

A 14-year-old boy presented with an 8 weeks history of occasional vomiting, weight loss, listlessness and increasing pallor.

During this period he complained intermittently of headache, pain in the lower chest anteriorly, and episodes of feeling hot and breathless. He had been short of breath on exertion. He had been drinking more water and passing more urine than previously. He complained of pains in his hands and feet and his family doctor arranged for an x-ray (Q9). His parents reported that since the onset of the illness his heart rate had become rapid and his heart beat unduly forceful.

He had a long history of episodes of fever, abdominal pain and vomiting which had been diagnosed as “abdominal migraine”. Both parents and his 4-year-old brother were healthy. His father was a factory worker and the family lived in a modern two-bedroomed flat.

On examination his weight was 30kg and his height was 138cm (growth charts Q11). He was alert and afebrile. His respiratory rate was 40/minute and his pulse rate was 130/minute. There was some pitting oedema over the dorsum of each foot. Jugular venous pressure was 5cm above the sternal angle. The apex beat was in the fifth interspace in the anterior axillary line and was thrusting in character. The first and second heart sounds were normal; the third heart sound was heard in the apical and left parasternal regions. The femoral pulses were readily palpable.

The blood pressure was 160/110 mmHg. Fine crepitations were heard at both lung bases. The appearance of the fundus is shown (Q10). The liver edge was palpable 3cm below the costal margin. Neither bladder nor kidneys could be palpated and there was no abdominal tenderness. Urinalysis was positive for protein (+) and negative for both glucose and blood.

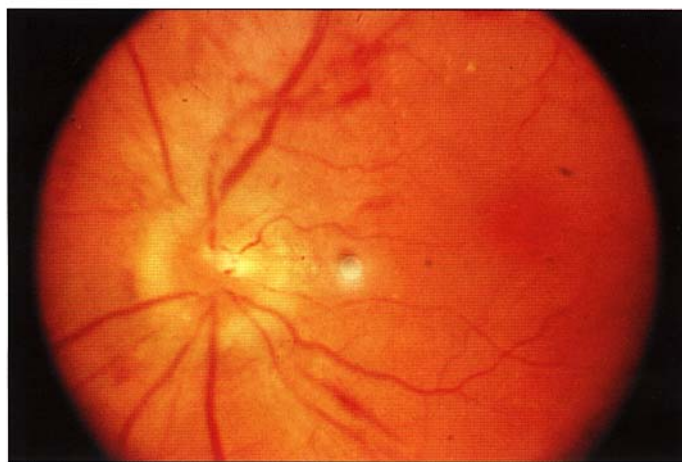
This question continues on the next page.

Hb	9.2 g/dl
MCV	73 fl
MCH	23 pg
MCHC	31 g/l
WBC	$8.0 \times 10^9/l$
neutrophils	$5.20 \times 10^9/l$
lymphocytes	$2.64 \times 10^9/l$
monocytes	$0.08 \times 10^9/l$
eosinophils	$0.08 \times 10^9/l$
plasma sodium	133 mmol/l
plasma potassium	4.0 mmol/l
plasma chloride	97 mmol/l
plasma bicarbonate	20 mmol/l
plasma urea	18 mmol/l
plasma creatinine	300 μ mol/l
plasma total protein	70 g/l
plasma albumin	38 g/l
plasma calcium	2.1 mmol/l
plasma phosphate	2.7 mmol/l (normal range 0.99-1.57)
plasma alkaline phosphate	496 IU/l (normal range for age 71-234)
Chest x-ray	normal
Abdominal ultrasound:	Kidneys small with increased echogenicity No bladder abnormality

This question continues on the next page.

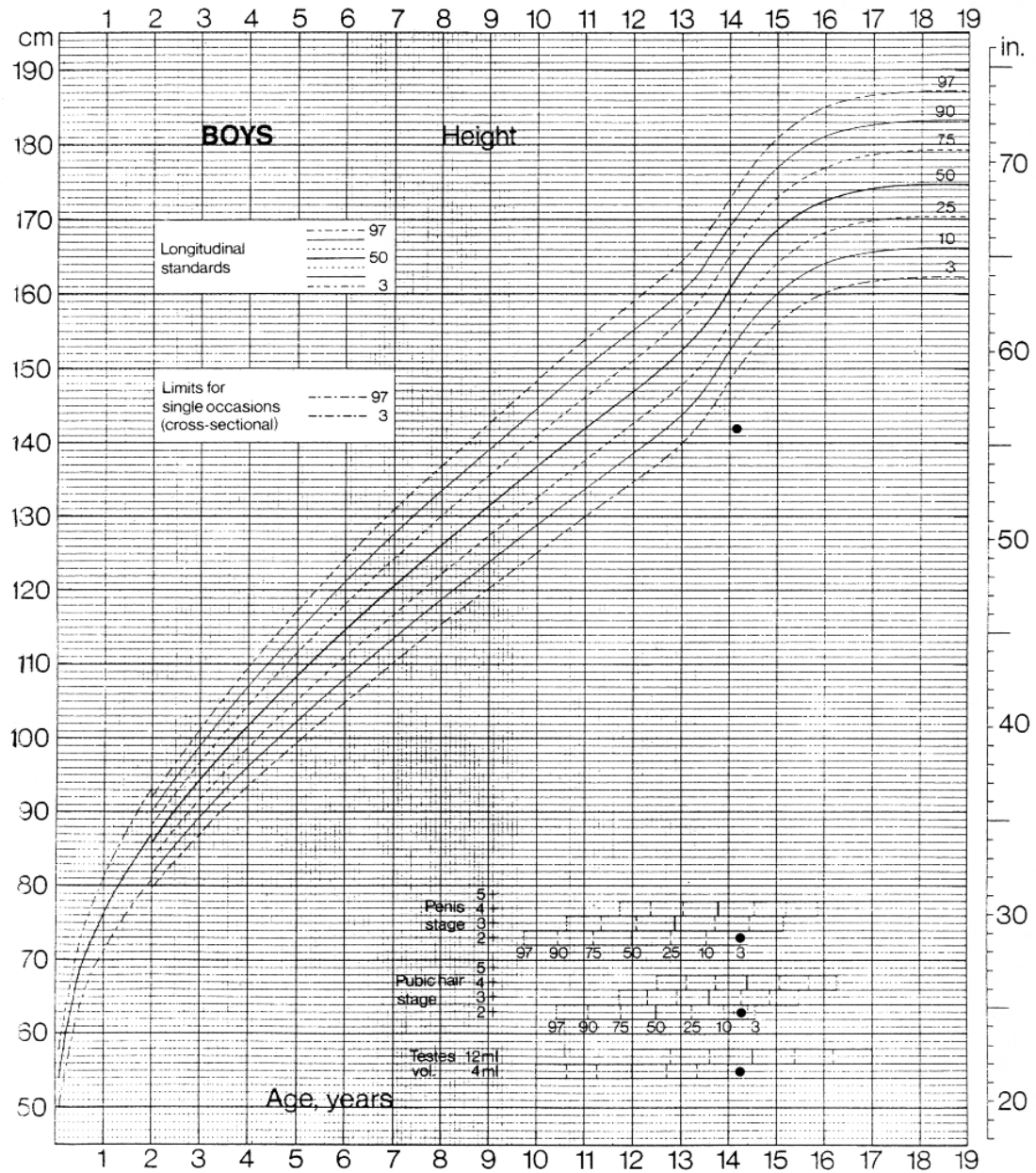


Q9



Q10

Q11



9. What is the most important abnormality on the radiograph of the hand shown in Q9? (1 mark)

SELECT ONE ANSWER ONLY

- A delayed bone age
- B osteomalacia
- C osteoporosis
- D splayed epiphyses
- E subperiosteal erosions

10. What is the most likely pathogenesis of the abnormality shown in Q9? (2 marks)

SELECT ONE ANSWER ONLY

- A chronic ill health
- B hypophosphataemia
- C poor dietary calcium intake
- D primary hyperparathyroidism
- E secondary hyperparathyroidism
- F vitamin D deficiency

This question continues on the next page.

11. What are the two most important features demonstrated on the growth chart? (3 marks)
(Q11)

SELECT TWO ANSWERS ONLY

- A bone age: advanced
- B bone age: delayed
- C bone age: normal
- D height: high
- E height: low
- F height: normal
- G pubertal staging: advanced
- H pubertal staging: delayed
- I pubertal staging: normal
- J weight for height: high
- K weight for height: low
- L weight for height: normal

This question continues on the next page.

- 12.** What is the best interpretation of the appearance of the optic fundus (Q12)? (3 marks)
(Ignore the central white spot which is an artefact)

SELECT ONE ANSWER ONLY

- A diabetic retinopathy
- B grade IV hypertensive retinopathy
- C optic atrophy
- D optic nerve head Drusen
- E pseudopapilloedema
- F traumatic retinal haemorrhage

- 13.** What is the most likely cause of his breathlessness? (2 marks)

SELECT ONE ANSWER ONLY

- A anaemia
- B left ventricular failure
- C metabolic acidosis
- D myocardial ischaemia
- E raised intracranial pressure
- F right ventricular failure

This question continues on the next page.

14. What is the most likely cause of his renal impairment? (2 marks)

SELECT ONE ANSWER ONLY

- A acute tubular necrosis
- B chronic glomerulonephritis
- C hypertensive nephropathy
- D hypovolaemia
- E reflux nephropathy

15. Which of the following renal investigations should now be performed? (2 marks)

SELECT TWO ANSWERS ONLY

- A abdominal CT
- B DMSA isotope scan
- C MAG 3 isotope scan
- D micturating cysto-urethrogram (MCUG)
- E renal arteriogram
- F renal biopsy

Questions 16, 17 and 18

- A acute lymphoblastic leukemia
- B beta thalassaemia major
- C beta thalassaemia trait
- D coeliac disease
- E congenital spherocytosis
- F Crohn's disease
- G haemolytic uraemic syndrome
- H immune haemolytic anaemia
- I Meckel's diverticulum
- J tuberculosis

Choose the one most likely diagnosis from the list above which best matches each of the following cases. **Note:** Each option may be used once, more than once or not at all.

SELECT ONE ANSWER ONLY FOR EACH QUESTION

- 16.** A previously well 2-year-old presents with a 3 week history of pallor, jaundice, lethargy and on examination has a spleen of 2 cms below the costal margin. The GP's full blood count shows Hb: 2.9gms white cell count $1.2 \times 10^9/l$, platelets 76, red cells indices normal. Lymphocytes 0.7, neutrophils 0.4.
- 17.** A normally grown 4-year-old Caucasian boy presents with lethargy and breathlessness of several weeks duration. On examination apart from pallor there is nothing else remarkable. Initial investigations have shown Hb 4 gm/dl. White cell count $7.0 \times 10^9/l$, platelets $520 \times 10^9/l$, reticulocytes 0.3%, blood film hypochromic/microcytic. Occasional atypical lymphocyte. ESR 25, ferritin 3.
- 18.** A 4-year-old presents with a 2-week history of lethargy and pallor following a viral illness. He looks slight jaundiced. Investigations show a Hb of 6.4 gms/dl, white cell count $12.5 \times 10^9/l$ (Neutrophils 5, lymphocytes 7, red cells 0.5), platelets $210 \times 10^9/l$. Blood film – MCV 90, occasional spherocytes, some atypical lymphocytes, no blast cells, reticulocytes count 6 %. Mycoplasma, IgM Positive.

Question 19

This is the face of a child aged three months with a normal platelet count who was otherwise well.

19. What is the best description of this lesion (3 marks)

SELECT ONE ANSWER ONLY

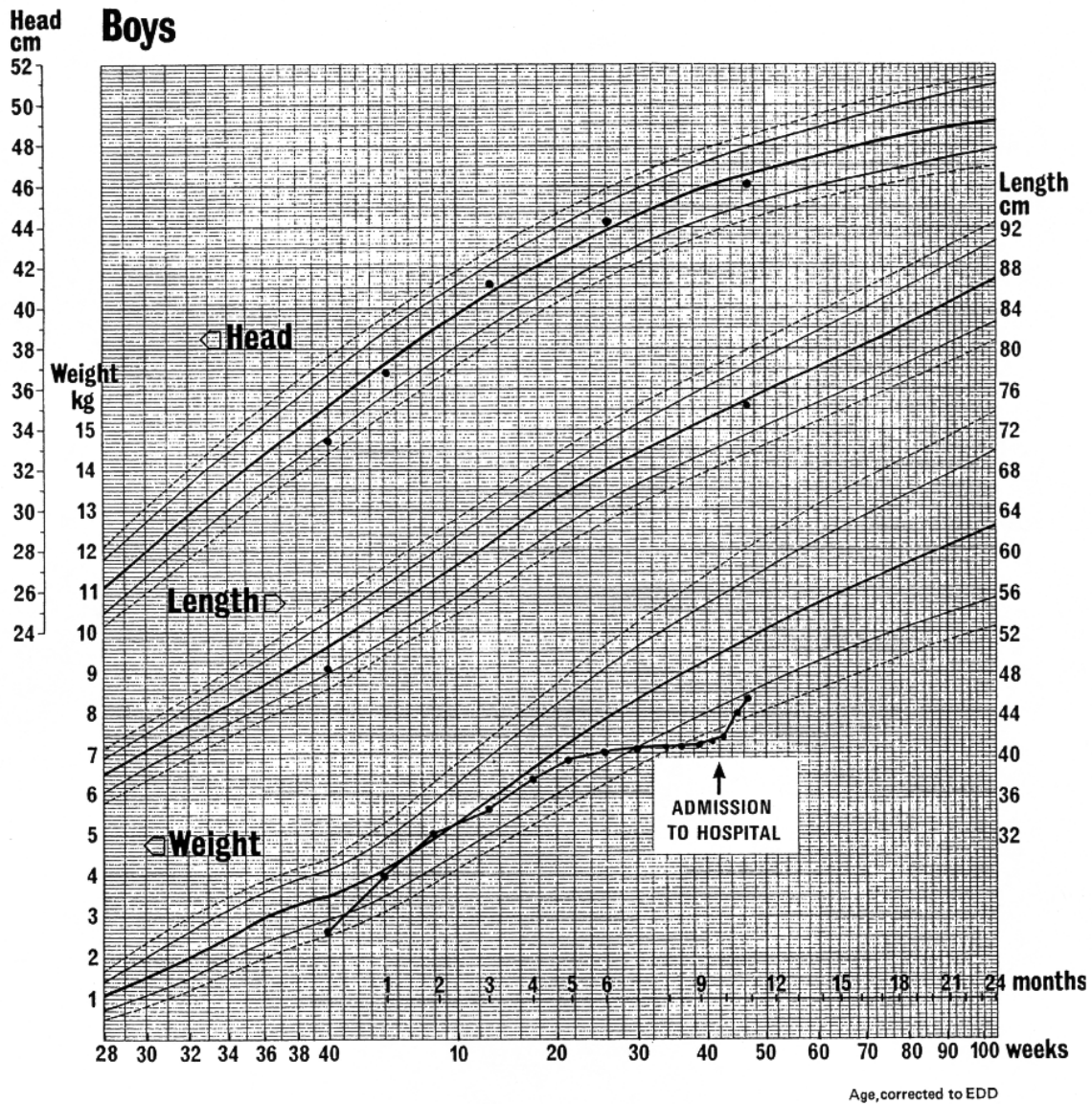
- A cavernous haemangioma
- B haemangioma
- C Kasabach-Merrit syndrome
- D mixed capillary/cavernous haemangioma
- E port wine naevus
- F cystic hygroma
- G strawberry naevus



Q19

Questions 20 and 21

This is the growth chart of a 10-month-old boy who was admitted to hospital because of vomiting and poor weight gain. The baby fed well on a mixed diet. The stools were normal. There were no abnormal findings apart from wasting.



Q20 & 21

This question continues on the next page.

20. What is the most likely diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A coeliac disease
- B cow's milk protein intolerance
- C cystic fibrosis
- D gastro-oesophageal reflux disease
- E neglect

21. In the light of this growth chart, select the single most helpful assessment. (2 marks)

SELECT ONE ANSWER ONLY

- A case conference
- B measure antigliadin and anti-endomysial antibodies
- C oesophageal pH monitoring
- D RAST (radioallergosorbent test) for cow's milk IgE
- E sweat test

Questions 22 and 23

The following haematological picture was found in a girl aged 16 years in early pregnancy. She was born in the UK to parents of Afro Caribbean origin, who have been very supportive of her during this unexpected pregnancy. She has no previous medical history of note.

Hb	10.5 g/dl
RBC	$5.50 \times 10^{12}/l$
PCV	0.32
MCV	58 fl
MCH	20 pg
WBC	$9.2 \times 10^9/l$ (normal differential)
Platelet count	$250 \times 10^9/l$

Red cell appearances microcytic and hypochromic; many target cells

22. Select one further investigation which would elucidate the cause of the anaemia (2 marks)

SELECT ONE ANSWER ONLY

- A ferritin
- B haemoglobin electrophoresis
- C red cell folate level
- D bone marrow aspiration
- E serum B12 level

This question continues on the next page.

23. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A anaemia due to folate deficiency
- B beta thalassaemia
- C beta thalassaemia trait
- D chronic blood loss
- E iron deficient anaemia
- F normal variation

Question 24

The illustration shows the tongue of an otherwise healthy child. Her mother was worried about this appearance.

24. Which of the following is the most appropriate management? (3 marks)

SELECT ONE ANSWER ONLY

- A biopsy of the white area
- B determine HIV status
- C follow up for observation
- D nystatin
- E offer reassurance
- F serum iron/ferritin



Q24

Question 25

This is the ECG of a 15-year-old who had surgery for congenital heart disease in childhood. He was asymptomatic apart from breathlessness on vigorous exertion.



Q25

25. What two abnormalities are shown on this ECG? (4 marks)

SELECT TWO ANSWERS ONLY

- A complete left bundle branch block
- B complete right bundle branch block
- C dextrocardia
- D first degree heart block
- E inferoposterior myocardial infarction
- F left axis deviation
- G left ventricular hypertrophy and strain
- H partial left bundle branch block
- I right ventricular hypertrophy and strain
- J Wolff-Parkinson-White syndrome

Question 26

A 10-month-old boy presented with a history of failure to gain weight, constipation, polydipsia and polyuria.

Investigations:

blood glucose	4.8	mmol/l
plasma urea	2.7	mmol/l
plasma creatinine	54	µmol/l
plasma sodium	129	mmol/l
plasma potassium	2.6	mmol/l
plasma chloride	101	mmol/l
plasma bicarbonate	18	mmol/l
plasma calcium	2.59	mmol/l
plasma phosphate	0.85	mmol/l (1.1-1.9 mmol/l)
plasma alkaline phosphatase	270	IU/l (normal range 71-212 IU/l)
plasma amino acids	All quantitatively normal	
pH	7.36	(hydrogen ion concentration 44 nmol/l)
Base deficit	-8.2	mmol/l

Urine analysis (24 hour collection)

volume	753mls on fluid intake 965mls	
pH	7.0	
glucose	12	mmol/l
sodium	25	mmol/l
potassium	16	mmol/l
urea	60	mmol/l
creatinine	0.4	µmol/l
phosphate	6.64	mmol/l
calcium	0.5	mmol/l
gross generalised amino-aciduria		
Ultrasound of kidneys and urinary tract	normal	
X-ray of skeleton	slight demineralisation but no other abnormality	

This question continues on the next page.

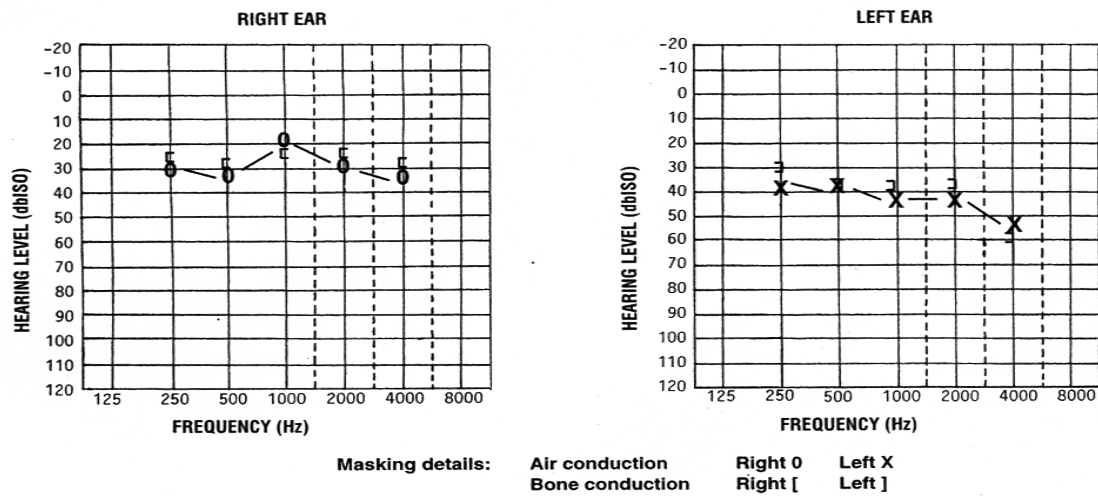
26. Choose three disturbances of renal physiology other than the amino-aciduria which have been demonstrated by these data. (4 marks)

SELECT THREE ANSWERS ONLY

- A acute renal failure
- B chronic renal failure
- C diabetes mellitus
- D excess hydrogen ion excretion
- E excess phosphate excretion
- F excess sodium excretion
- G inadequate phosphate excretion
- H inadequate potassium excretion
- I inadequate sodium excretion
- J renal glycosuria

Question 27

This is the audiogram of a 7-year-old child.



Q27

27. What abnormality is revealed by this audiogram? (5 marks)

SELECT ONE ANSWER ONLY

- A bilateral moderate conductive hearing loss
- B bilateral moderate sensorineural hearing loss
- C bilateral severe conductive hearing loss
- D bilateral severe sensorineural hearing loss
- E left moderate conductive hearing loss
- F left moderate sensorineural hearing loss
- G left severe conductive hearing loss
- H left severe sensorineural hearing loss
- I right moderate conductive hearing loss
- J right moderate sensorineural hearing loss
- K right severe conductive hearing loss
- L right severe sensorineural hearing loss

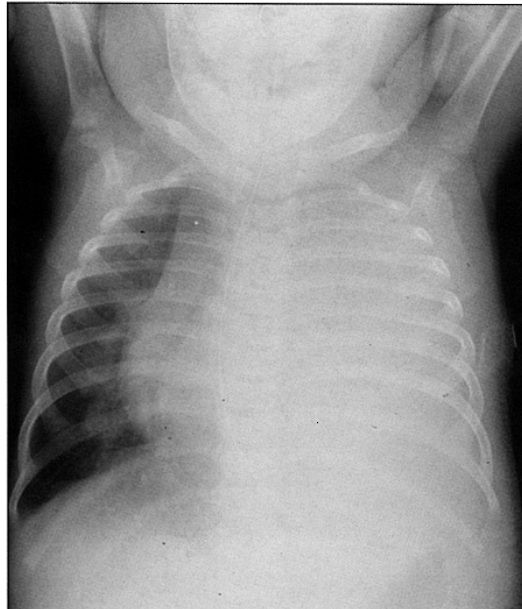
Question 28

This 2-year-old boy presented with a two weeks history of cough, fever and increasing breathlessness.

28. What are the two most important abnormalities present? (4 marks)

SELECT TWO ANSWERS ONLY

- A left lower lobe consolidation
- B left pleural effusion
- C left upper and lower lobe consolidation
- D left upper lobe consolidation
- E mediastinal shift
- F right lower lobe consolidation
- G right middle lobe consolidation
- H right pleural effusion
- I right upper lobe consolidation
- J right widespread consolidation



Q28

Questions 29, 30 and 31

A	ACTH	I	Lorazepam
B	Carbamazepine	J	Paraldehyde
C	Clonazepam	K	Phenobarbitone
D	Diazepam	L	Phenytoin
E	Ethosuximide	M	Sodium valproate
F	Gabapentin	N	Topiramate
G	Ketogenic diet	O	Vigabatrin
H	lamotrigine	P	None

The above drugs are used for the treatment of epilepsy.

Which drug would you use as a first line treatment in the following cases?

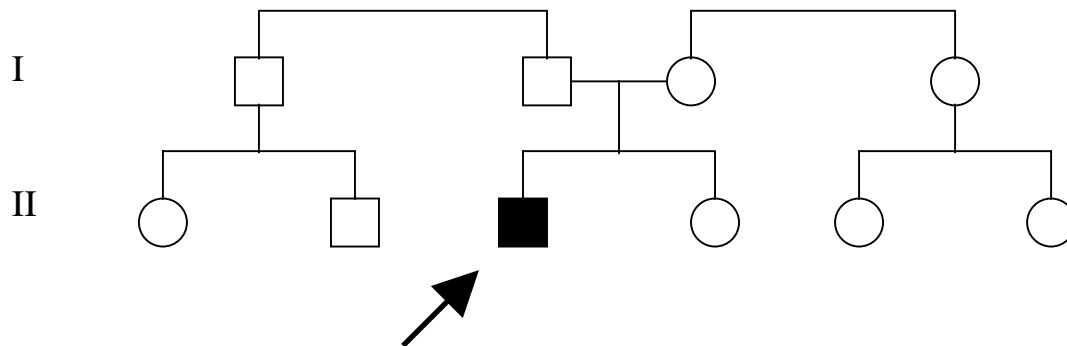
Note: *Each option may be used once, more than once or not at all.*

SELECT ONE ANSWER ONLY FOR EACH QUESTION

- 29.** A term newborn with birth asphyxia presents at 12 hours of age with frequent short-lived generalised convulsions.
- 30.** A 16-year-old girl doing her GCSE exams with generalised tonic epilepsy.
- 31.** A 7-year-old boy with generalised absence seizures.

Questions 32 and 33

This index child possesses the karyotype 47, XXY. His parents, aged 25 and 26, have normal karyotypes.



32. What is the degree of risk of his parents having subsequent children with this condition? (2 marks)

SELECT ONE ANSWER ONLY

- A zero
- B less than 1%
- C 1%
- D 5%
- E 25%
- F 50%
- G 67%
- H 100%

This question continues on the next page.

33. What is the likelihood of the index child having affected offspring? (2 marks)

SELECT ONE ANSWER ONLY

- A zero
- B less than 1%
- C 1%
- D 5%
- E 25%
- F 50%
- G 67%
- H 100%

Question 34

34. This is the face of a boy aged five years.

What is the most likely diagnosis? (3marks)

SELECT ONE ANSWER ONLY

- A eczema herpeticum
- B herpes simplex
- C impetigo
- D Stevens Johnson syndrome
- E toxic epidermal necrolysis



Q34

END OF SPECIMEN PAPER 1

Paper One Web Specimen Paper - Answer Key			
Question Number	Answer Key	Answer and Description	Marks
1	B	nasopharyngeal aspirate for respiratory viruses	3
2	F	viral bronchiolitis	4
3	A	left sided Horner's syndrome	3
4	B	partially compensated respiratory acidosis	2
5	B	ensure nasal CPAP circuit is working correctly	3
6	C	CT scan	3
7	E	administer intravenous mannitol	2
	G	arrange for intubation and ventilation	2
8	E	subarachnoid haemorrhage	3
9	E	subperiosteal erosions	1
10	E	secondary hyperparathyroidism	2
11	E	Height: low	1
	H	pubertal staging: delayed	2
12	B	grade IV hypertensive retinopathy	3
13	B	left ventricular failure	2
14	E	reflux nephropathy	2
15	B	DMSA isotope scan	1
	D	micturating cysto-urethrogram (MCUG)	1
16	A	acute lymphoblastic leukaemia	3
17	F	Crohn's disease	3
18	H	immune haemolytic anaemia	3
19	A	cavernous haemangioma	3
20	E	neglect	2
21	A	case conference	2
22	B	haemoglobin electrophoresis	3
23	C	beta thalassaemia trait	2
24	E	offer reassurance	3
25	D	first degree heart block	2
	I	right ventricular hypertrophy and strain	2
26	E	excess phosphate excretion	1
	F	excess sodium excretion	1
	J	renal glycosuria	1
27	B	bilateral moderate sensorineural hearing loss	5
28	B	left pleural effusion	2
	E	mediastinal shift	2
29	K	Phenobarbitone	3
30	H	Lamotrigine	3
31	M	Sodium valproate	3
32	B	less than 1%	2
33	A	zero	2
34	D	Stevens Johnson syndrome	3

CANDIDATE NUMBER:.....

Royal College of Paediatrics and Child Health
MRCPCH PART 2 EXAMINATION

PAPER 1

1. Complete the following:

Your full name (BLOCK LETTERS)

.....

RCPCH Number

Signature

2. Please check that your surname (family name) is correct on the **Answer Sheet**.
3. Using the pencil provided, answer all of the following 60 questions. **Do not** write anything on the Answer Sheet other than your response to each question.
4. Where questions are based upon photographs, X-rays, growth-charts, ECGs, or any other visual material, the question will refer you to the **Clinical Photographs Booklet**.
5. It is strictly forbidden to talk to, read the work of, or attempt in any way to communicate with, other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work. The College has tools which can identify copying of answers or collusion between candidates to share answers. In any situation the suspicion of guilt falls upon both parties until it can be proved otherwise. Breaches of these instructions, or misbehaviour in any other way, including continuing to write after the allotted time, may lead to suspension from the examination at the discretion of the invigilators. Serious breaches, such as cheating or colluding to gain advantage, could incur permanent suspension from College examinations.
6. Copyright law protects examination questions and the intellectual property of their authors. The unauthorised use of questions is a breach of copyright law.
7. Time allowed: 2 hours 30 minutes.

HOW TO COMPLETE THE ANSWER SHEET

The answer sheet contains a row of boxes for each question. Each box contains a letter. Indicate your answer(s) by filling in the appropriate box(es) with the pencil provided. The number of answers to be selected is stated in the question. If more than the stated number of answers are chosen, **no** marks will be awarded for the question. If you choose fewer answers than the stated number, marks will be given for each correct choice. The three types of question formats used in this examination are:

Best of List

In this question you are asked to choose the best single answer from a list.

Example 1

1. What is the most likely diagnosis?

SELECT ONE ANSWER ONLY

- A glucose 6 phosphate dehydrogenase deficiency.
- B haemophilia A.
- C haemorrhagic disease of the newborn.
- D hereditary spherocytosis.
- E Von Willebrand's disease.

If you choose B this would be indicated on the answer sheet as:

1. cA > ☒ cB cC cD cE cF cG cH > cI > cJ cK > cL cM cN cO cP > cQ >

Choose n from many

In this question you are asked to choose a number of options from a longer list. For example the signs on a chest X-ray, or the findings on an ECG.

Example 2

2. What three investigations are most likely to be helpful in the next few hours?

SELECT THREE ANSWERS ONLY

- A abdominal ultrasound.
- B blood culture.
- C blood glucose.
- D C reactive protein (CRP).
- E capillary blood gas analysis.
- F chest X-ray.
- G CT head.
- H electroencephalogram (EEG).
- I full blood count.
- J lumbar puncture.
- K plain abdominal X-ray.
- L referral for surgical opinion.
- M urea, creatinine and electrolytes.
- N urine culture.

If you choose B, G and J this would be indicated on the answer sheet as:

2 cA > ☒ cB cC cD cE cF ☒ cG cH > cI > ☒ cJ cK > cL cM cN cO cP > cQ >

Extended Matching

In this question, a list of options is given at the beginning of the question. This might include a list of diagnoses, treatments, drugs or other management steps. After the list, short clinical scenarios or statements are made. You will be asked to choose the option(s) in the list which is(are) most appropriate.

Example 3

- A carbamazepine.
- B clonazepam.
- C ethosuximide.
- D gabapentin.
- E lamotrigine.
- F phenytoin.
- G prednisolone.
- H topiramate.
- I valproate.
- J vigabatrin.

Questions 3, 4 and 5

Choose the most appropriate drug from the list above in each of the following situations (Note: Each option may be used once, more than once or not at all):

SELECT ONE ANSWER ONLY FOR EACH SITUATION

Note: Each answer can be used more than once

3. five month-old child presents to outpatients. Her mother tells you . . .
4. an eight year-old boy presents with episodes of collapse at school His teacher...
5. a five year-old girl has a long history of epilepsy. She has previously been treated...

If you choose J, I and D respectively, this would be indicated on the answer sheet as:

3	<input type="radio"/> A	<input type="radio"/> B	<input type="radio"/> C	<input type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input type="radio"/> G	<input type="radio"/> H	<input type="radio"/> I	<input checked="" type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
.	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input checked="" type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
4	<input type="radio"/> A	<input type="radio"/> B	<input type="radio"/> C	<input type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input type="radio"/> G	<input type="radio"/> H	<input type="radio"/> I	<input type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
.	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
5	<input type="radio"/> A	<input type="radio"/> B	<input type="radio"/> C	<input checked="" type="radio"/> D	<input type="radio"/> E	<input type="radio"/> F	<input type="radio"/> G	<input type="radio"/> H	<input type="radio"/> I	<input type="radio"/> J	<input type="radio"/> K	<input type="radio"/> L	<input type="radio"/> M	<input type="radio"/> N	<input type="radio"/> O	<input type="radio"/> P
.	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

General Instructions

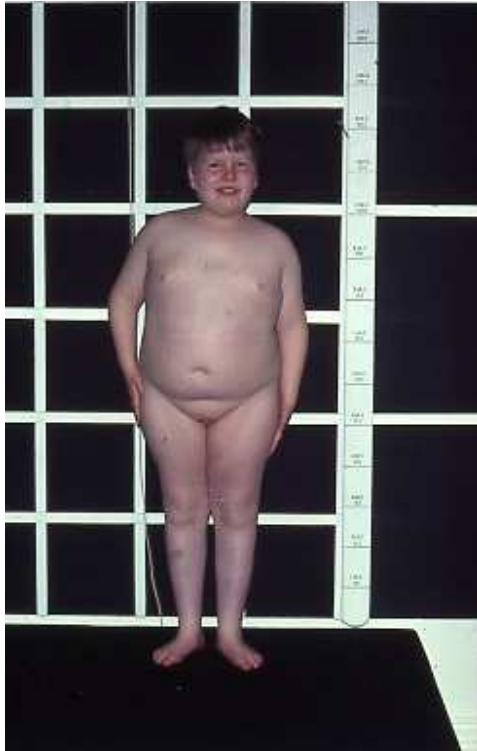
1. Do not fold or crease the answer sheet or make any marks except in the spaces provided.
2. It is recommended that initially you indicate your answer against the questions in the question book. You should leave sufficient time (say half an hour) to transfer your answers to the answer sheet.
3. Answer the question by placing a neat bold horizontal line across the letter of your choice within the box.
4. The scoring machine has been programmed to ignore erasures (which can leave smudges) and this means that faint responses or partial responses may be ignored by the machine.
5. Smudges due to erasures may be caused by dirty rubbers - cleaning can be accomplished by rubbing against a clean part of the question book. Let the invigilator know if you are worried about the appearance of your erasures.

BLANK PAGE

Please do not turn over this page until instructed to do so by the invigilator.

Questions 1 and 2

This 10 year-old boy is referred to outpatients because of poor school performance and behavioural problems. Educational assessment suggests mild learning difficulty. Blood pressure and blood glucose are normal. His height is on the 9th centile and weight >99.6th centile.



1. Which investigation is most likely to lead to a diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A chromosomal analysis
- B cortisol profile
- C pituitary function tests
- D serum calcium and parathormone levels
- E thyroid function tests

This question continues on the next page.

2. What are the next most appropriate actions? (4 marks)

SELECT TWO ANSWERS ONLY

- A dietitian opinion
- B educational psychology report
- C measure height: leg length ratio
- D measure parents' body mass index
- E pubertal assessment
- F refer for exercise programme
- G referral to clinical geneticist
- H referral to endocrinologist
- I x-ray for bone age

Questions 3, 4 and 5

The following is a list of cardiac diagnoses

- A aortic regurgitation
- B aortic stenosis
- C atrial septal defect
- D coarctation of the aorta
- E mitral regurgitation
- F mitral stenosis
- G patent arterial duct (ductus arteriosus)
- H pulmonary stenosis
- I transposition of the great arteries
- J ventricular septal defect

For each of the following clinical scenarios select the most likely diagnosis

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

3. A 2 year-old boy presents with an asymptomatic murmur found on routine examination of the chest for an intercurrent chest infection. The peripheral pulses were normal but an ejection systolic murmur was heard over the right upper sternal area which radiated to the neck and along the left sternal edge. A systolic thrill was palpable in the suprasternal notch. (3 marks)
4. A 2 year-old boy presents with an asymptomatic murmur found on routine examination of the chest for an intercurrent chest infection. There was an ejection systolic murmur and a thrill palpable in the left upper sternal area. The first and second heart sounds were clearly separated. (3 marks)
5. A 2 year-old girl presents with an asymptomatic murmur found on routine examination of the chest for an intercurrent chest infection. There was a blowing systolic murmur best heard at the apex and radiating to the axilla. An apical low pitched systolic murmur was also heard. The second heart sound was increased but not split. (3 marks)

Question 6

An 8 year-old boy presents to outpatients having recently noticed the development of pubic hair. He is having difficulty at school and is disruptive in class. He has a history of asthma for which he is taking inhaled fluticasone at the recommended dose for age.

On examination there are no abnormalities detected apart from pubic hair, his testicular volumes are 2 mls bilaterally.

Investigations

Blood

sodium	128 mmol/l
potassium	5.8 mmol/l
bicarbonate	22 mmol/l
urea	3.4 mmol/l
creatinine	62 µmol/l

6. Which of the following is the most likely enzyme deficiency in his case? (4 marks)

SELECT ONE ANSWER ONLY

- A 11-beta hydroxylase
- B 17 alpha-hydroxylase
- C 21-hydroxylase
- D 3 alpha reductase
- E 3 beta-hydroxy-steroid dehydrogenase

Question 7

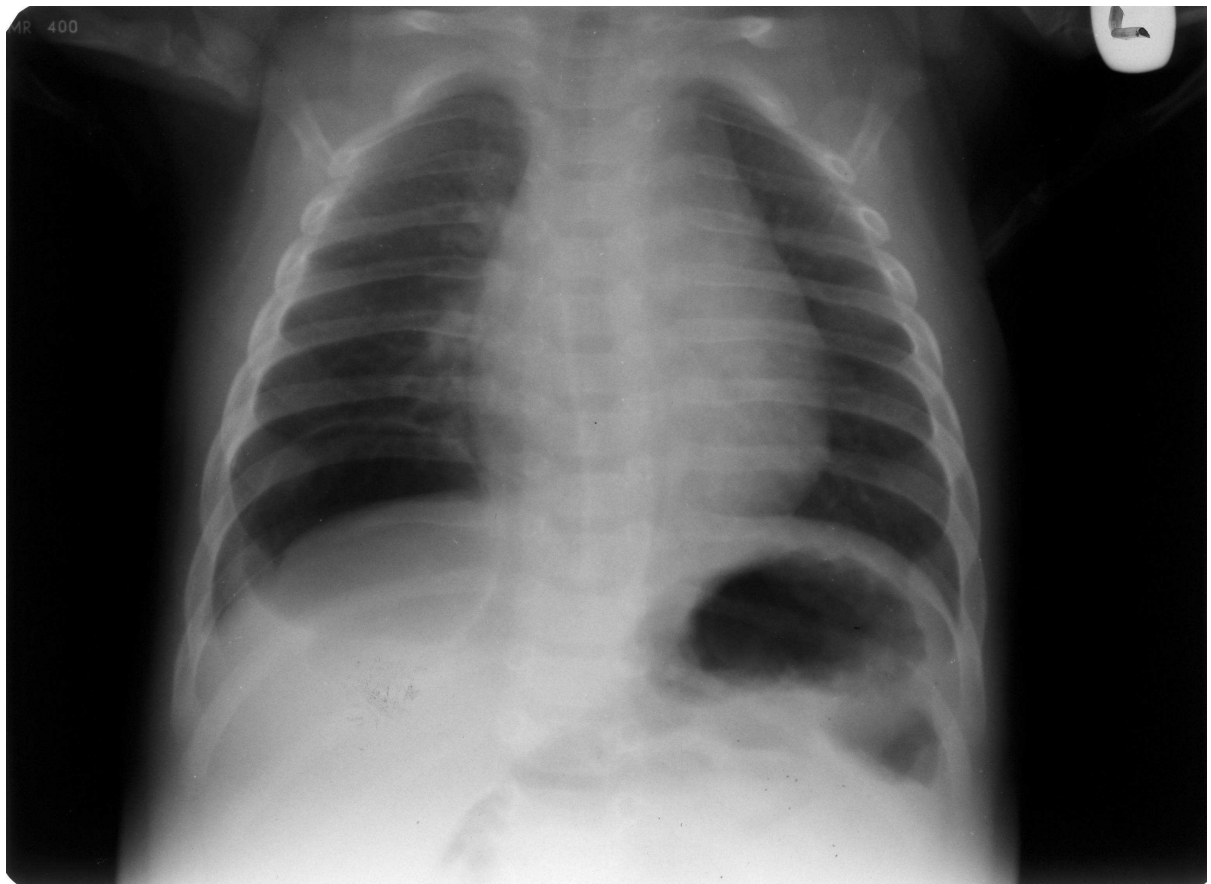
7. Which two of the following types of fractures would make you particularly suspicious of physical abuse? (4 marks)

SELECT TWO ANSWERS ONLY

- A complete fracture of the clavicle in a 3 year-old
- B fracture of the femur in an ambulant child
- C fracture of the tibia in an ambulant child
- D posterior rib fracture under one year of age
- E spiral fracture of the humerus in a non-ambulant child
- F spiral fracture of the humerus in an ambulant child

Question 8

An 18 month-old boy presented with history of persistent cough for 4 weeks. He has had an URTI 3 weeks previously but had recovered with only symptomatic treatment. On examination his chest was clear to auscultation, his heart sounds were normal and his abdomen was soft with no organomegaly present. His chest x-ray is shown in the figure below.

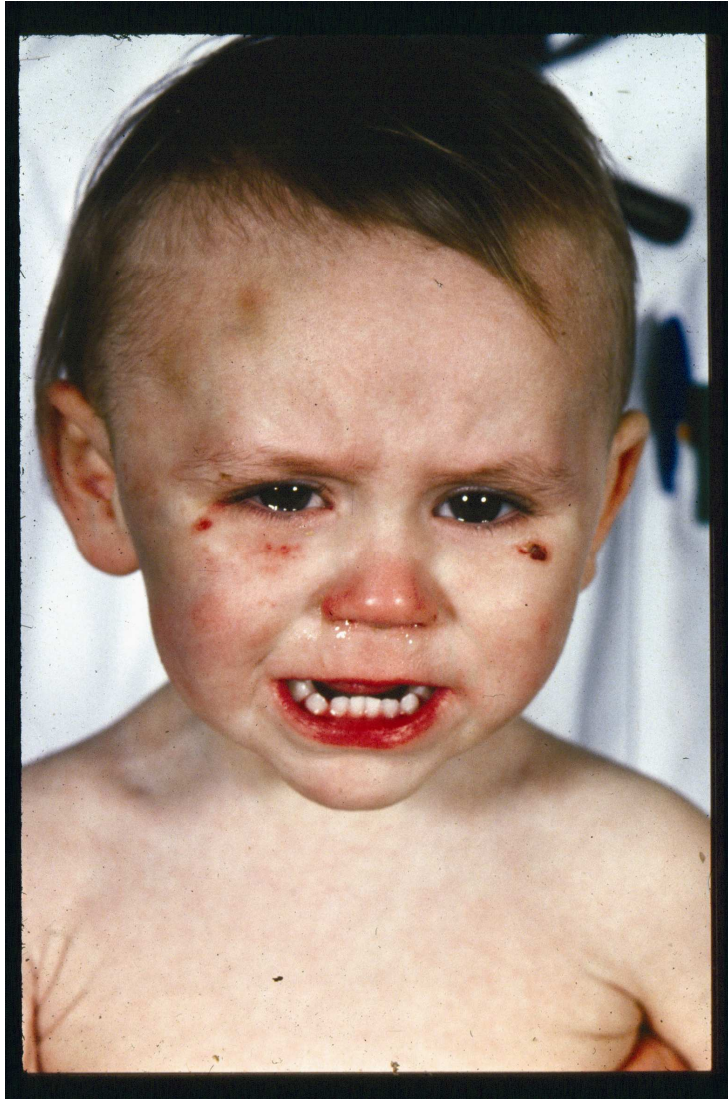


8. What is the most likely diagnosis? (4 marks)

- A congenital diaphragmatic hernia
- B congenital lobar emphysema
- C congenital lung cyst
- D loculated pneumothorax
- E pneumatocele

Question 9

9. What is the most likely cause of the skin lesions on this child's face? (3 marks)



SELECT ONE ANSWER ONLY

- A accidental injury
- B acute lymphoblastic leukaemia
- C haemangiomas
- D idiopathic thrombocytopenic purpura injury
- E non-accidental injury

Questions 10, 11 and 12

The following is a list of anti-inflammatory drugs

- A anti-TNF alpha
- B azathioprine
- C ciclosporin
- D cyclophosphamide
- E intravenous immunoglobulin
- F oral prednisolone
- G methotrexate
- H rituximab
- I sulphasalazine
- J tacrolimus

Which of the above medications is most likely to be the cause of each of the following clinical scenarios?

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: *Each answer may be used more than once*

- 10. 7 year old boy with severe asthma and multiple hospital admissions has reduced growth velocity in the last 6 months. (3 marks)
- 11. A 16 year-old girl with steroid resistant Crohn's disease develops neutropenia on alternative treatment. (3 marks)
- 12. A 5 year-old boy 3 weeks post BMT for relapsed ALL develops macroscopic haematuria. (3 marks)

Question 13

A 14 year-old girl is referred to outpatients with an 18 month history of fatigue and recurrent abdominal pain.

On examination her height is 142 cm (0.4th centile) and weight 37 kg (2nd centile).

Investigations

Blood

haemoglobin	10.1 g/dl
MCH	26 pg
MCV	70 fl
white cell count	$9.6 \times 10^9/l$
platelets	$260 \times 10^9/l$
ferritin	6 $\mu g/l$ (15-300)
CRP	9 mg/l

13. Which of the following would be the most useful investigation? (4 marks)

- A bone age
- B colonoscopy and biopsy
- C IgA anti-tissue transglutaminase antibodies
- D immunoglobulins
- E karyotype
- F upper g-i contrast study

Question 14

A 14 month-old boy has had one confirmed Coliform urinary tract infection. He was treated with a 7-day course of oral antibiotics and is now well.

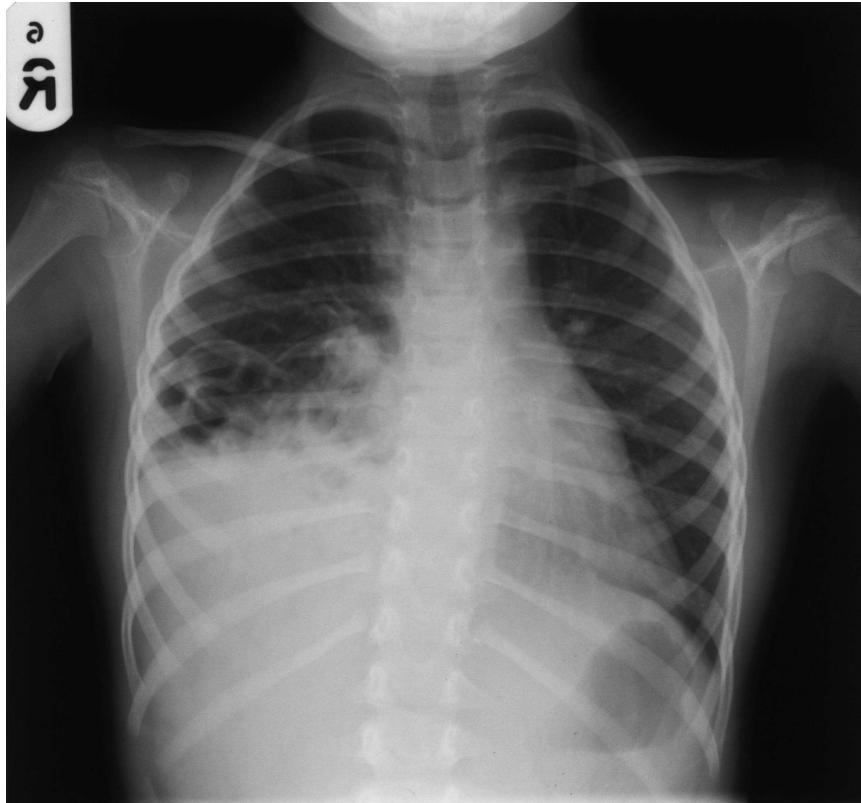
14. Which of the following investigations is most appropriate at this stage? (3 marks)

SELECT ONE ANSWER ONLY

- A DMSA scan
- B micturating cystogram
- C ultrasound scan of renal tract
- D all of the above
- E none of the above

Question 15

This is the chest x-ray of an 18 month-old girl who presented with a history of cough, fever and noisy breathing for the previous 48 hours. She has previously been well and her height and weight were on the 25 to 50th centiles for age



15. Which of the following is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A bronchiectasis
- B congenital cystic adenomatoid malformation
- C diaphragmatic hernia
- D foreign body aspiration
- E pneumatoceles
- F primary ciliary dyskinesia
- G pulmonary tuberculosis

Question 16

This infant was referred from the midwifery led unit having been born at term following an uneventful pregnancy. His clinical photograph is shown below.



16. What is the most important initial investigation? (4 marks)

SELECT ONE ANSWER ONLY

- A chromosome analysis
- B cranial CT scan
- C cranial ultrasound
- D EEG
- E full blood count
- F intraocular pressure measurement

Question 17, 18 and 19

The following is a list of treatment options for seizures

- A carbamazepine
- B lamotrigine
- C levetiracetam
- D no treatment
- E phenobarbitone
- F phenytoin
- G sodium valproate
- H topiramate
- I vigabatrin
- J zonisamide

Choose the most appropriate treatment for each of the following cases

SELECT ONE ANSWER ONLY

Note: *Each answer may be used more than once*

- 17. A 13 year-old girl with juvenile absence epilepsy. (3 marks)
- 18. A 6 month-old boy with infantile spasms and developmental delay. (3 marks)
- 19. An 8 year-old boy with infrequent nocturnal seizures and bilateral centrottemporal spikes on EEG. (3 marks)

Questions 20 and 21

This 7 month-old boy presents with a 4-day history of rash associated with low grade fever, cough and rhinitis. The rash started as erythematous papules over the trunk. He is irritable and has not been feeding well. Nikolsky's sign is positive.



20. What is the most appropriate diagnostic step? (3 marks)

- A detailed drug history
- B Epstein-Barr virus serology
- C HIV serology
- D Mycoplasma serology
- E nasal swab for culture
- F skin biopsy

21. What is the intravenous medication of choice? (3 marks)

- A AZT
- B erythromycin
- C flucloxacillin
- D fluconazole
- E immunoglobulin
- F methylprednisolone

Question 22

A 4 month old baby boy from Eastern Europe who has recently immigrated to the UK presents with poor feeding, loose stools and faltering growth. On examination his length and weight are on the 2nd centiles for age. He is noted to have oral candidiasis a mild degree of bilateral parotid swelling.

Investigations

Blood

haemoglobin	8.3 g/dl
white cell count	$5.2 \times 10^9/l$
neutrophils	$3.1 \times 10^9/l$
lymphocytes	$2.0 \times 10^9/l$
platelets	$485 \times 10^9/l$
immunoglobulins	IgG 9.5 g/dl (2.1-7.7)
	IgA 1.2 g/dl (0.05-0.4)
	IgM 1.8 g/dl (0.15-0.7)
chest x-ray	bilateral patchy infiltrates

22. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A chronic granulomatous disease
- B common variable immune deficiency
- C HIV infection
- D severe combined immune deficiency
- E X-linked hypogammaglobulinaemia

Question 23

This 9 month-old male infant presents with a 3-day history of fever, rhinorrhoea and rash. His mother had brought him to see the GP, who had prescribed erythromycin and antipyretics. The child did not improve and his rash worsened extending from his face to the rest of his body, in particular the axillae and groins.



23. What is the diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A atopic dermatitis
- B Herpes simplex infection with secondary bacterial infection
- C impetigo
- D Staphylococcal scalded skin syndrome
- E Stevens-Johnson syndrome
- F toxic epidermal necrolysis

Question 24

A 15 year-old girl with focal epilepsy comes to clinic having had 5 seizures in the past 2 weeks. Her seizures were previously well-controlled on carbamazepine. She was otherwise well except for a cough over the past month for which she has been taking erythromycin. She has recently started on the oral contraceptive pill.

24. What would be the next step with management? (3 marks)

SELECT ONE ANSWER ONLY

- A Add other anti-epileptic drug
- B Check carbamazepine level
- C Consider another form of contraception
- D CT head scan
- E Increase dose of carbamazepine

Question 25

This 9 month-old infant was cyanosed at birth and had a cardiac operation at 3 months of age.



25. What condition is shown here? (3 marks)

SELECT ONE ANSWER ONLY

- A left sided Horner's syndrome
- B left sided lower motor neurone facial palsy
- C left sided ptosis
- D left sided upper motor neurone facial palsy
- E right-sided Horner's syndrome
- F right-sided lower motor neurone facial palsy
- G right-sided ptosis
- H right-sided upper motor neurone facial palsy

Question 26

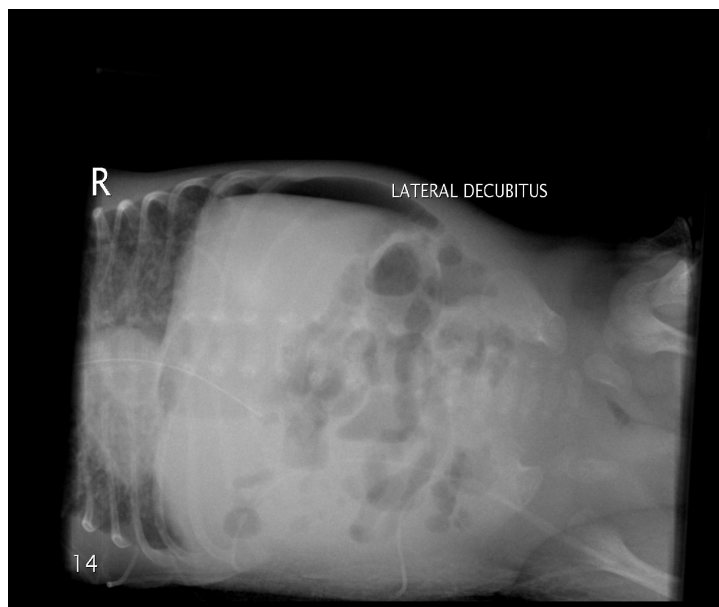
This 28-week gestation baby, currently ventilated, has had a worsening metabolic acidosis over the last 24 hours. His heart rate is 180/minute and mean blood pressure is 26 mmHg. He has developed peripheral oedema. Due to bilious aspirates an abdominal x-ray was performed.

Investigations

Blood gas

pH	7.01 (hydrogen ion 100mmol/l)
pCO ₂	5.5kPa (41.2 mmHg)
pO ₂	8.1kPa (61 mmHg)
HCO ₃	10.2 mmol/l
BE	-17.0 mmol/l

His abdominal x-ray



26. What is the most appropriate immediate management? (4 marks)

SELECT ONE ANSWER ONLY

- A administer 1mmol/kg sodium bicarbonate
- B fluid resuscitation with 10mls/kg 0.9% saline
- C increase ventilation pressures and rate
- D needle aspiration
- E start intravenous antibiotics

Question 27

A 12 year-old boy with learning disability was seen in the child development centre with a 12-month history of increasingly awkward gait and difficulty getting footwear. On examination he has a scoliosis, marked pes cavus, absent lower limb reflexes and extensor plantar responses.

27. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A dermatomyositis
- B Duchenne muscular dystrophy
- C Friedreich's ataxia
- D hereditary motor and sensory neuropathy
- E spinal cord tumour

Question 28

A 12 year-old boy was admitted with a 2 day history of fever, vomiting and diarrhoea. His parents were from Singapore. He had recently returned from a trip to Malaysia. Physical examination revealed a slightly lethargic looking boy with dry mucus membranes, but otherwise comfortable with good peripheral perfusion. Cardio-respiratory examination was unremarkable. The abdomen was not distended or tender, his spleen was palpable at 3 cm below the left costal margin. He was commenced on intravenous rehydration.

Investigations

Blood

haemoglobin	12.8 g/dl
MCV	56.8 fl
white cell count	$4.8 \times 10^9/l$
platelets	$210 \times 10^9/l$

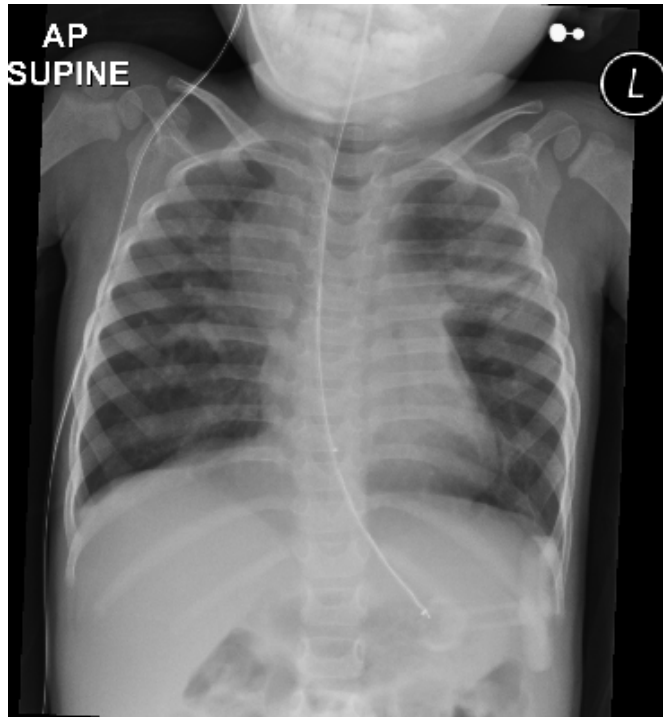
28. What is the most appropriate investigation to confirm the cause of the splenomegaly?
(3 marks)

SELECT ONE ANSWER ONLY

- A blood film examination for malarial parasites
- B bone marrow aspirate
- C haemoglobin electrophoresis
- D osmotic fragility test
- E serum ferritin
- F glucose-6-phosphate dehydrogenase level

Questions 29 and 30

A 20 month-old girl has recurrent cough, intermittent vomiting and failure to thrive. She had been born at 29 weeks gestation and was oxygen dependent till 10 months of age. Her chest x-ray is shown in the figure below.



29. Which of the following investigations is most appropriate at this stage? (4 marks)

SELECT ONE ANSWER ONLY

- A Bronchoscopy
- B CT thorax
- C Echocardiography
- D serum immunoglobulins
- E sputum culture
- F video-swallow

This question continues on the next page.

30. What treatment should this girl be receiving? (3 marks)

SELECT ONE ANSWER ONLY

- A antibiotics
- B anti-reflux therapy
- C anti-viral agents
- D chemotherapy
- E removal of thoracic mass
- F no treatment required

Question 31

This is the face of a child aged 3 months with a normal platelet count who is otherwise well.



31. What is the best description of this lesion? (3 marks)

SELECT ONE ANSWER ONLY

- A cavernous haemangioma
- B haemangioma
- C Kasabach-Merrit syndrome
- D mixed capillary/cavernous haemangioma
- E port wine naevus
- F cystic hygroma
- G strawberry naevus

Questions 32, 33 and 34

The following is a list of endocrine diagnoses

- A constitutional growth and pubertal delay
- B growth hormone deficiency
- C hypothalamic hamartoma
- D idiopathic precocious puberty
- E Klinefelter syndrome
- F McCune – Albright syndrome
- G premature adrenarche
- H premature thelarche
- I testicular tumour
- J Turner syndrome

For each of the following clinical scenarios choose the most likely diagnosis.

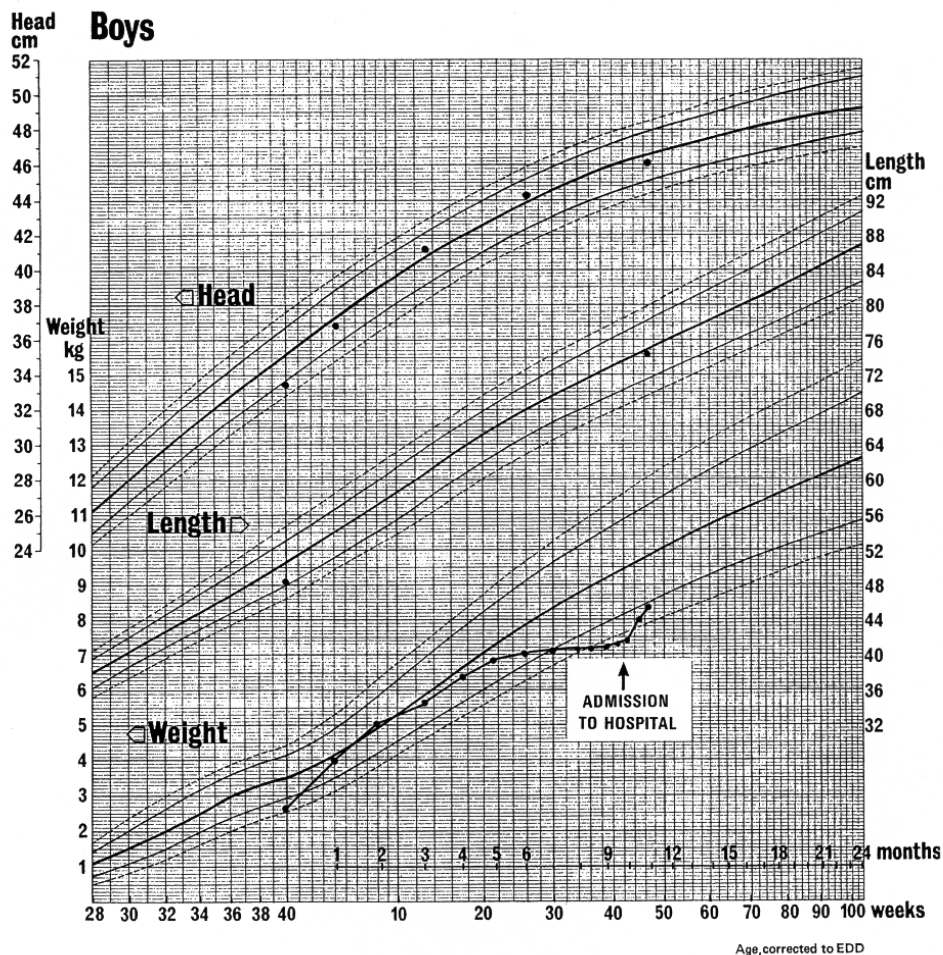
SELECT ONE ANSWER ONLY

Note: *Each answer may be used more than once*

- 32. A 2 year-old girl with isolated bilateral breast development. (3 marks)
- 33. A 7 year-old boy with axillary and pubic hair development and testicular volume of 2 mls. (3 marks)
- 34. A 14 year-old boy with short stature and a family history of delayed puberty. (3 marks)

Questions 35 and 36

This is the growth chart of a 10 month-old boy who was admitted to hospital because of vomiting and poor weight gain. The baby fed well on a mixed diet. The stools were normal. There were no abnormal findings apart from wasting.



This question continues on the next page.

35. What is the most likely diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A coeliac disease
- B cow's milk protein intolerance
- C cystic fibrosis
- D gastro-oesophageal reflux disease
- E inadequate calorie intake

36. In the light of this growth chart, select the single most helpful next action. (2 marks)

SELECT ONE ANSWER ONLY

- A discharge planning meeting
- B measure anti-tTG antibodies
- C oesophageal pH monitoring
- D RAST test for cow's milk IgE
- E sweat test

Question 37

Study: Moderate Hypothermia to Treat Perinatal Asphyxial Encephalopathy. **Background:** It is uncertain whether hypothermic therapy improves neurodevelopmental outcomes in newborn infants with asphyxial encephalopathy. **Methods:** A trial was performed in infants less than 6 hours old and at least 36 weeks gestation with hypoxic encephalopathy. The trial compared intensive care plus cooling of the body to 33.5°C for 72 hours and intensive care alone. The primary outcome was death or severe disability at 18 months of age. **Results:** Of 325 infants enrolled after obtaining permission from parents, 163 underwent intensive care with cooling, and 162 underwent intensive care alone. In the cooled group, 42 infants died and 32 survived but had severe neurodevelopmental disability, whereas in the non-cooled group, 44 infants died and 42 had severe disability (relative risk for either outcome, 0.86; (95% confidence interval [CI] 0.68 to 1.07). Relative risk (RR) of survival without neurologic abnormality in the cooled group was 1.57 (95% CI, 1.16 to 2.12). Among survivors, cooling resulted in cerebral palsy with RR of 0.67 (95% CI, 0.47 to 0.96).
Adapted from: NEJM 2009;361:1349-1358

37. Which one of the following best describes this study: (3 marks)

SELECT ONE ANSWER ONLY

- A case control study
- B meta-analysis
- C observational study
- D prospective cohort study
- E randomised controlled trial

Question 38

A 3 day-old baby girl was admitted with poor feeding, recurrent apneas and poor peripheral pulses. She required resuscitation and ventilatory support. Arterial blood gases showed a severe metabolic acidosis. She responded well to prostacycline infusion and oxygen saturation increased from 84% to 95%.

What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A interrupted aortic arch
- B sepsis
- C transposition of great arteries
- D tricuspid atresia
- E urea cycle disorder

Question 39

A 4 week-old baby boy is seen in outpatients with a history of sudden jerky movements noticed by his mother almost from birth. They are seen most prominently whilst the baby is asleep. His mother thinks they are more obvious recently and may number from one or two to about ten or more in a few hours sleep. Each jerk seems to last a few seconds and appears to involve an isolated limb. Physical examination is normal.

Which of the following is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A benign sleep myoclonus
- B drug withdrawal syndrome
- C hypocalcaemia
- D infantile spasms
- E myoclonic epilepsy

Question 40

A 4 year-old child presents to outpatients with his mother, father and elder brother. He has been referred with a pansystolic murmur at the apex heard by a GP at a pre-school medical. During the course of the examination he is noted to be on the 90th centile for height and the 25th centile for weight and has an asymmetrical chest. He has hypermobile joints and a history of joint pains.

40. Select the two most appropriate initial clinical responses to the scenario. (4 marks)

SELECT TWO ANSWERS ONLY

- A an orthopaedic opinion
- B ASO titre
- C chromosomal markers for Marfan's Syndrome
- D echocardiography
- E enquire about a family history of sudden death
- F examine family members for similar characteristics

Question 41

Randomised controlled trial of nasal continuous positive airways pressure (CPAP) in bronchiolitis.

Aims: To compare CPAP with standard treatment (ST) in the management of bronchiolitis.

Methods: Children <1 year of age with bronchiolitis and a capillary $\text{PCO}_2 > 6 \text{ kPa}$ were recruited and randomised to CPAP or ST and crossed over to the alternative treatment after 12 hours. ST was intravenous fluids and supplemental oxygen by nasal prongs or face mask. The change in PCO_2 was compared between the groups after 12 and 24 hours. A secondary outcome was the need for invasive ventilatory support.

Results: 29 of 31 children completed the study. PCO_2 after 12 hours fell by 1.35 kPa (95% confidence interval (CI) 2.03 to 0.67 kPa) in children treated with CPAP first compared to a 0.53 kPa (95% CI 1.1 to -0.05) for those on ST. No children in the CPAP group but 2 in the standard treatment group required ventilation.

Adapted from Arch Dis Ch 2007; 93:45-47

41. Which of the following conclusions can be drawn from this data? (6 marks)

SELECT TWO ANSWERS ONLY

- A A double-blind cross-over trial would be a more practical way of testing this hypothesis
- B CPAP reduces the need for invasive ventilation in children with bronchiolitis
- C CPAP should be the standard treatment for infants with bronchiolitis
- D Measured FiO_2 would have been a better measure of response to treatment
- E Paired t-test is the most appropriate statistical method for the analysis of the changes in PCO_2 obtained in the study
- F The cross-over design actually used increases the power of the study

Question 42

An 18 month-old girl is seen in her local community-based clinic. She is from a family who travel the country with her father, a labourer. Her grandmother recently died of tuberculosis. The family were seen and the child's chest x-ray was normal. One week ago she was given BCG immunisation. She has had a cough and nasal discharge for 1 to 2 weeks. She appears well and her temperature is 37.2°C.

She has had no other immunisations. The health visitor notes a high non-attendance rate and points out the need to make progress with her immunisation programme.

42. Which of the following is the most appropriate course of action? (6 marks)

SELECT ONE ANSWER ONLY

- A Bring back in one week to give diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines.
- B Bring back in six weeks to give diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines.
- C Offer diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines and measles, mumps and rubella immunisation immediately.
- D Offer diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines immediately.
- E Offer measles, mumps and rubella immunisation alone immediately.

Questions 43, 44 and 45

The following is a list of respiratory diagnosis

- A bronchiectasis
- B Chlamydia pneumonitis
- C gastro-oesophageal reflux
- D hyper IgE syndrome
- E obliterative bronchiolitis
- F pertussis
- G primary ciliary dyskinesia
- H pulmonary tuberculosis
- I recurrent aspiration
- J tracheo-oesophageal fistula

For each of the following clinical scenarios choose the most likely diagnosis from the list above.

SELECT ONE ANSWER ONLY FOR EACH QUESTION

Note: Each answer may be used more than once

- 43. A 5 year-old girl with recurrent otitis media and chronic rhinorrhoea who presents with a persistent cough. Crackles are heard in the right mid-zone. (3 marks)
- 44. A 2-year old boy who suffered from severe birth asphyxia presents with recurrent cough and wheeze. He had mild eczema as an infant. (3 marks)
- 45. A 3 year-old boy with a history of recurrent skin sepsis, eczema, chronic cough and sputum production. Sputum samples grow Staphylococcus aureus. (3 marks)

Questions 46 and 47

A 14 month-old boy is admitted with rotavirus gastroenteritis. He has a history of tetralogy of Fallot with a right Blalock-Taussig shunt. On examination he was pale and tachypnoeic with a capillary return of 5 seconds. First and second heart sounds were heard and a continuous murmur was present over the right infra-clavicular area. A review of the nursing chart showed that he was on milk feeds and has opened his bowels 13 times today.

Investigations

Capillary blood gas

pH	7.16 (hydrogen ion concentration 70 nmol/l)
PCO ₂	3.9 k Pa (29.5 mmHg)
pO ₂	4.0. kPa (31mmHg))
BE	-17 mmol/l
HCO ₃	10.4 mmol/l

46. What is the best description of the acid base abnormalities? (3 marks)

SELECT ONE ANSWER ONLY

- A fully compensated metabolic acidosis
- B fully compensated metabolic alkalosis
- C fully compensated respiratory acidosis
- D fully compensated respiratory alkalosis
- E partially compensated metabolic acidosis
- F partially compensated respiratory alkalosis

This question continues on the next page.

47. What is the next most appropriate step in management? (3 marks)

SELECT ONE ANSWER ONLY

- A 10ml/kg of normal saline
- B 2ml/kg of 4.2% sodium bicarbonate over 1 hour
- C intravenous isoprenaline
- D intravenous morphine
- E intravenous propranolol
- F put the child in the knee chest position

Questions 48 and 49

A female infant of 32 weeks gestation, on her 4th day of life, was well maintained on intermittent mandatory ventilation at 10 breaths per minute. She was changed to nasal CPAP in 40% oxygen at 5cm water pressure.

One hour after commencing nasal CPAP the following results were obtained from an arterial blood gas.

pH	7.20 (hydrogen ion concentration 62 nmol/l)
PaCO ₂	10.0 kPa (75mmHg)
PaO ₂	11.7 kPa (88mmHg)
base excess	1.1 mmol/l
standard bicarbonate	28 mmol/l

48. Select the best description for the acid-based abnormality. (2 marks)

SELECT ONE ANSWER ONLY

- A compensated metabolic acidosis
- B compensated respiratory acidosis
- C normal
- D partially compensated metabolic acidosis
- E partially compensated respiratory acidosis
- F uncompensated metabolic acidosis
- G uncompensated respiratory acidosis

49. What would be your next step in the management of this infant? (3 marks)

SELECT ONE ANSWER ONLY

- A chest x-ray
- B ensure nasal CPAP circuit is operating correctly
- C increase inspired oxygen concentration
- D reintubate and restart ventilation
- E transilluminate chest

Question 50

A 15 year-old boy with a diagnosis of asthma for 10 years was referred by his GP to outpatients because of persisting respiratory symptoms. At the time of referral he was on inhaled beclometasone 1 milligram twice daily and inhaled salmeterol 50 micrograms twice daily for the last six months. He is also on inhaled salbutamol 2-10 puffs as required. His Peak Expiratory Flow rate was 255 L/minute (71% predicted). He was not able to participate fully in school sports.

50. Which two of the following investigations would you do next? (3 marks)

SELECT TWO ANSWERS ONLY

- A bronchial lavage
- B bronchoscopy
- C chest x-ray
- D Mantoux test
- E nasopharyngeal brushings
- F short synacthen test
- G spiral CT of chest
- H sweat test
- I ventilation/perfusion lung scan

Question 51

A previously well 16 year-old boy presented to outpatients with a 6-month history of frequent headaches. Recent problems with balance had occurred while playing football for his school team. The headaches were relieved by lying flat. There was a strong family history of migraine.

Physical examination was normal apart from a positive Romberg's sign and unsteadiness on attempting to heel-toe walk. Reflexes were normal.

51. Which of the following is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A Arnold-Chiari malformation type 1
- B idiopathic (benign) intracranial hypertension
- C late onset muscular dystrophy (Becker)
- D migraine
- E spinal cord tumour

Question 52

A 10 year-old boy with type 1 diabetes since the age of 8 comes to clinic for his annual review. His growth is normal for age, his control has been excellent and his HbA1c is 6.3%.

52. Which of the following conditions known to be associated with diabetes should be screened for at this age? (4 marks)

SELECT TWO ANSWERS ONLY

- A coeliac disease
- B hypercholesterolaemia
- C hypothyroidism
- D nephropathy
- E peripheral neuropathy
- F retinopathy

Question 53

A 12 year-old girl presented to the outpatient department with a 3-month history of headache, morning vomiting and weight loss. Her mother had died in a road traffic accident one year previously. She had been attending a psychiatrist for symptoms of depression since that time.

On examination she was thin and mildly dehydrated. Apart from apparent difficulty with bilateral upward gaze her neurological examination was normal with no abnormality of limb power, tone or reflexes. There was no papilloedema and her blood pressure was 110/65 mmHg.

53. Which of the following is the most likely finding on the cranial MRI? (4 marks)

SELECT ONE ANSWER ONLY

- A benign intracranial hypertension
- B herniation of the medulla
- C intracranial tumour
- D mid-brain haemorrhage causing superior oblique muscle paralysis (Parinaud syndrome)
- E normal scan
- F subarachnoid haemorrhage

Question 54

A 4 year-old child presents with his fifth febrile seizure. He is developmentally normal and his father had a history of febrile seizures as a child.

54. Which of the following is most appropriate at this stage? (2 marks)

SELECT ONE ANSWER ONLY

- A CT head scan
- B ECG
- C EEG
- D MRI brain scan
- E no investigation

Question 55

A 10 month-old boy presents to the emergency department at 7am after being found at home unrousable in his cot. Nursing staff note a few bruises over his forehead and limbs. He was born at term and spent three weeks on the neonatal unit with neonatal abstinence syndrome. His mother reports him to have been hot the previous night. On examination he is breathing comfortably and his oxygen saturations are 95% in room air. His Glasgow Coma Scale score is 13, his capillary refill is time 3-4 seconds.

55. What is the most appropriate initial investigation? (3 marks)

SELECT ONE ANSWER ONLY

- A blood gas
- B blood glucose
- C fundoscopy
- D lumbar puncture
- E urgent CT head scan

Question 56

A 2 year-old Afro-Caribbean girl was referred with concern about her locomotor development. Further history taking revealed that she was largely breast fed and was described as a “fussy eater”. She lived with her mother and new boyfriend in a high rise council block. Her height and weight were 2nd to 9th centiles for age. Her wrist x-ray is shown in figure below.



56. Which of the following is the most likely explanation for her symptoms? (3 marks)

SELECT ONE ANSWER ONLY

- A metaphyseal dysplasia
- B mucopolysaccharidosis
- C non-accidental injury
- D osteogenesis imperfecta
- E rickets
- F sickle cell anaemia

Question 57

A 9 month-old boy presented to the emergency department with a 24 hour history of irritability and decreased movements of his left arm. His parents said that he had accidentally rolled off the couch onto the floor the previous evening. His x-ray is shown in figure below.



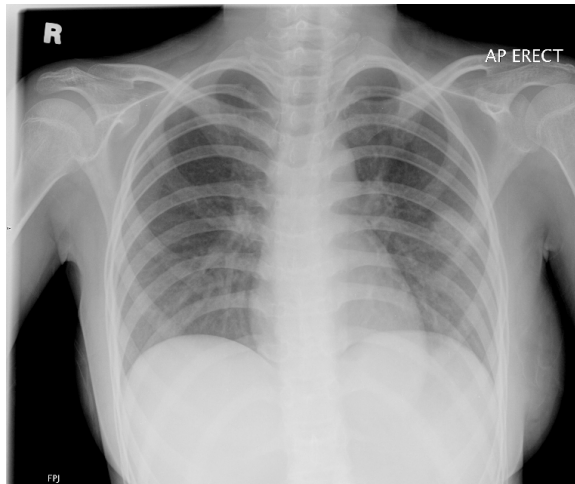
57. What is the most likely cause of this appearance? (3 marks)

SELECT ONE ANSWER ONLY

- A accidental injury
- B non-accidental injury
- C osteogenesis imperfecta
- C osteopenia
- E rickets

Question 58

A 12 year-old girl with frontal lobe epilepsy presents with a 4-day history of fever cough vomiting and drowsiness. Her chest x-ray is shown in the figure below.



Investigations

Blood

sodium	116 mmol/l
potassium	2.8 mmol/l
chloride	90 mmol/l
urea	1.8 mmol/l
creatinine	40 μ mol/l
bicarbonate	19 mmol/l
osmolality	247 mmol/kg

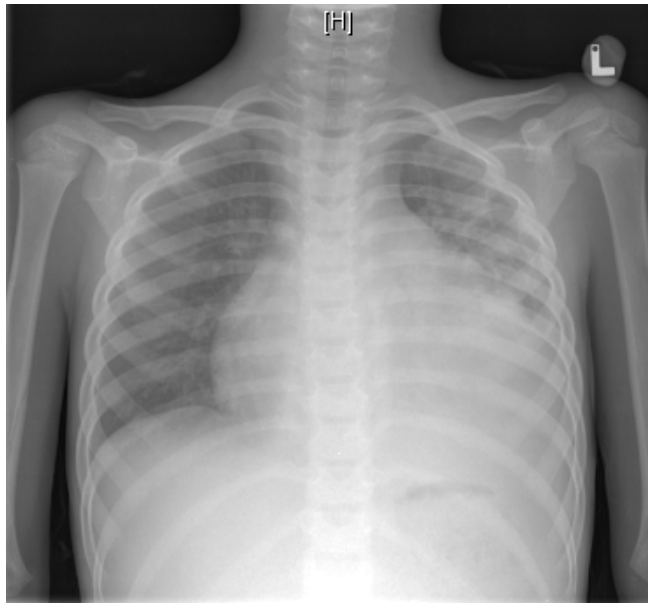
urine osmolality	<u>107</u> mmol/kg
sodium	18 mmol/l
potassium	19 mmol/l
chloride	< 15 mmol/l

58. Which of the following is the most likely cause of her current biochemical findings? (3 marks)

- A Addison's disease
- B cerebral salt wasting
- C excessive water drinking
- D pseudohyponatremia
- E syndrome of inappropriate anti-diuretic hormone secretion (SIADH)

Question 59 and 60

A 7 year-old boy presents with fever, cough, dyspnoea and generalized edema. His chest x-ray is shown in the figure below.



59. What is the most appropriate investigation to further establish the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A blood culture
- B CT thorax
- C echocardiography
- D Mycoplasma serology
- E pleural tap for culture
- F sputum culture

This question continues on the next page.

60. What is the most appropriate treatment? (3 marks)

SELECT ONE ANSWER ONLY

- A amoxycillin
- B chest tube drainage
- C erythromycin
- D furosemide
- E inhaled salbutamol
- F oral steroid

THE END



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MRCPCH PART 2 EXAMINATION

**Answer Key
PAPER 1**

Q No	Ans	Marks	Answer in words
1	A	3	chromosomal analysis
2	G	2	referral to clinical geneticist
	H	2	referral to endocrinologist
3	B	3	aortic stenosis
4	C	3	atrial septal defect
5	E	3	mitral regurgitation
6	C	4	21-hydroxylase
7	D	2	posterior rib fracture under one year of age
	E	2	spiral fracture of the humerus in a non-ambulant child
8	C	4	congenital lung cyst
9	E	3	non-accidental injury
10	F	3	Oral prednisolone
11	B	3	azathioprine
12	D	3	cyclophosphamide
13	C	4	anti-tTG antibodies
14	E	3	None of the above
15	C	4	Diaphragmatic hernia
16	F	4	intraocular pressure measurement
17	B	3	lamotrigine
18	I	3	vigabatrin
19	D	3	No treatment
20	E	3	nasal swab for culture
21	C	3	flucloxacillin
22	C	3	HIV infection
23	D	4	Staphylococcal scalded skin syndrome
24	C	3	Consider another form of contraception
25	A	3	left sided Horner's syndrome
26	B	4	Fluid resuscitation
27	C	3	Friedreich's Ataxia
28	C	3	haemoglobin electrophoresis
29	F	4	video-swallow
30	B	3	anti-reflux therapy
31	A	3	cavernous haemangioma
32	H	3	Premature thelarche
33	G	3	Premature adrenarche
34	A	3	Constitutional growth and pubertal delay
35	E	2	inadequate calorie intake
36	A	2	discharge planning meeting
37	E	3	randomised controlled trial
38	A	3	interrupted aortic arch
39	A	3	benign sleep myoclonus
40	D	2	echocardiography
	F	2	examine family members for similar characteristics



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MRCPCH PART 2 EXAMINATION

**Answer Key
PAPER 1**

Q No	Ans	Marks	Answer in words
41	E	3	Paired t-test is the most appropriate statistical method for the analysis of the changes in PCO ₂ obtained in the study
	F	3	The cross-over design actually used increases the power of the study
42	D	6	Offer diphtheria, tetanus, pertussis, polio, Haemophilus influenzae B, Meningitis C and Pneumococcal conjugate vaccines immediately.
43	G	3	primary ciliary dyskinesia
44	I	3	recurrent aspiration
45	D	3	hyper IgE syndrome
46	E	3	partially compensated metabolic acidosis
47	A	3	10ml/kg of normal saline
48	E	2	partially compensated respiratory acidosis
49	B	3	ensure nasal CPAP circuit is operating correctly
50	C	2	chest x-ray
	F	2	short synacthen test
51	A	4	Arnold-Chiari malformation type 1
52	A	4	coeliac disease
53	C	4	intracranial tumour
54	E	2	No investigation
55	B	3	blood glucose
56	E	3	rickets
57	B	3	non-accidental injury
58	C	3	excessive water drinking
59	C	3	echocardiography
60	D	3	furosemide
THE END			

Royal College of Paediatrics and Child Health
MRCPCH PART II EXAMINATION

PAPER 1

1. Complete the following:

Your full name (BLOCK LETTERS)

.....

RCPCH Number

Signature

.....

2. Please check that your surname (family name) is correct on the **Answer Sheet**.

Do not write anything on the Answer sheet other than your response to each question.

3. Answer all of the questions.

4. It is strictly forbidden to talk to, read the work of, or attempt in any way to communicate with, other candidates whilst the examination is in progress. Please exercise vigilance to ensure that no other candidate can attempt to copy your work. The College has tools which can identify copying of answers or collusion between candidates to share answers. In any situation the suspicion of guilt falls upon both parties until it can be proved otherwise. Breaches of these instructions, or misbehaviour in any other way, including continuing to write after the allotted time, may lead to suspension from the examination at the discretion of the invigilators. Serious breaches, such as cheating or colluding to gain advantage, could incur permanent suspension from College examinations.

5. Copyright law protects examination questions and the intellectual property of their authors. The unauthorised use of questions is a breach of copyright law.

6. Time allowed: 1 hour.

Questions 1 and 2

A 12-week-old baby was found by his parents blue and apparently lifeless in his cot after they had heard him choking. His father revived him with mouth-to-mouth resuscitation and brought him to the hospital.

He was born at 36 weeks gestation weighing 2.4 kg. He had no perinatal problems and fed well by bottle although he frequently regurgitated his feeds. His parents reported that he had been unwell with a cough for a few days before admission. His two siblings aged 5 years and 3 years had both been unwell recently with chest infections. Father had been unemployed for 18 months and there were considerable financial problems.

On examination he was pale and had a nasal discharge. His weight was on the 10th centile. Slight intercostal recession and a respiratory rate of 40/minute were noted together with scattered fine crepitations and a high pitched expiratory wheeze. The remainder of the examination was normal. Examination of the urine revealed no protein or sugar and no cells were seen on microscopy. A chest x-ray showed a degree of overinflation with no localised opacities.

1. What is the most useful investigation for establishing the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A echocardiogram
- B nasopharyngeal aspirate for respiratory viruses
- C pernasal swab
- D pH probe
- E sweat test

This question continues on the next page.

2. What is the most likely diagnosis? (4 marks)

SELECT ONE ANSWER ONLY

- A congenital cardiac abnormality
- B cystic fibrosis
- C gastro-oesophageal reflux
- D milk aspiration pneumonitis
- E pertussis infection
- F viral bronchiolitis

Question 3

This 9-month-old infant was cyanosed at birth and had a cardiac operation at 3 months of age.

3. What condition is shown here? (3 marks)

SELECT ONE ANSWER ONLY

- A left sided Horner's syndrome
- B left sided lower motor neurone facial palsy
- C left sided ptosis
- D left sided upper motor neurone facial palsy
- E right-sided Horner's syndrome
- F right-sided lower motor neurone facial palsy
- G right-sided ptosis
- H right-sided upper motor neurone facial palsy



Q3

Questions 4 and 5

An infant of 32 weeks gestation on his fourth day of life was well maintained on intermittent mandatory ventilation at 10 breaths per minute for respiratory distress syndrome. She was changed to nasal continuous positive airway pressure (CPAP) in 40% oxygen ($FiO_2 = 0.4$) at 5cm water pressure.

One hour after commencing nasal CPAP the following results were obtained from an arterial blood gas specimen:

pH	7.20 (hydrogen ion concentration 62 nmol/l)
PaCO ₂	10.0 kPa (75mmHg)
PaO ₂	11.7 kPa (88mmHg)
Base excess	1.1 mmol/l
Standard bicarbonate	24 mmol/l
Actual bicarbonate	28.8 mmol/l

4. Select the best description for the acid-based abnormality. (2 marks)

SELECT ONE ANSWER ONLY

- A compensated metabolic acidosis
- B compensated respiratory acidosis
- C normal
- D partially compensated metabolic acidosis
- E partially compensated respiratory acidosis
- F uncompensated metabolic acidosis
- G uncompensated respiratory acidosis

This question continues on the next page.

5. What would be your next step in the management of this infant? (3 marks)

SELECT ONE ANSWER ONLY

- A chest X-ray
- B ensure nasal CPAP circuit is operating correctly
- C increase inspired oxygen concentration
- D reintubate and restart ventilation
- E transilluminate chest

Questions 6, 7 and 8

A 14-year-old boy of Bangladeshi origin was seen in the Accident & Emergency Department of a District General Hospital with a generalised convulsion. His parents said that he had complained for two weeks previously of mild headaches, which had occurred at different times of the day. At the age of 12 he was found to be sniffing glue but subsequently told his parents he had discontinued the practice. His progress at school was good and his behaviour had been normal.

On the afternoon of admission he had complained of a sudden generalised headache; despite this he had gone to see some friends but returned home with the headache. His mother had given him paracetamol. As he was sitting down to watch television, he became stiff and had a generalised convulsion.

The family called an ambulance and rectal diazepam was administered. He continued to fit and on arrival at the hospital, intravenous lorazepam was required to terminate the convulsion. He remained very drowsy and non-responsive.

On examination, there was some resistance to flexion of his neck but he was afebrile. His respirations were laboured, he was not cyanosed and was well perfused peripherally. His blood pressure was 160/90 mmHg. Examination of his heart, respiratory system and abdomen were normal.

His pupils were of equal size and both reacted sluggishly to light. Examination of the fundi showed no abnormalities; there was a generalised increase in tone in his limbs but no focal abnormal neurological signs.

6. What is the most appropriate investigation to establish the diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A lumbar puncture
- B Mantoux test
- C CT scan
- D toxicology screen
- E urgent EEG

This question continues on the next page.

7. What are the two most appropriate forms of immediate management? (4 marks)

SELECT TWO ANSWERS ONLY

- A Administer intravenous antibiotics
- B Administer intravenous dexamethasone
- C Administer intravenous diuretic
- D Administer intravenous fluid bolus
- E Administer intravenous mannitol
- F Administer second-line anti-epileptic medication
- G Arrange for immediate intubation and ventilation
- H Discuss patient with a Paediatric Intensive Care Unit

8. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A cerebral tumour
- B drug overdose
- C epilepsy
- D meningitis
- E subarachnoid haemorrhage

Questions 9, 10, 11, 12, 13, 14 and 15

A 14-year-old boy presented with an 8 weeks history of occasional vomiting, weight loss, listlessness and increasing pallor.

During this period he complained intermittently of headache, pain in the lower chest anteriorly, and episodes of feeling hot and breathless. He had been short of breath on exertion. He had been drinking more water and passing more urine than previously. He complained of pains in his hands and feet and his family doctor arranged for an x-ray (Q9). His parents reported that since the onset of the illness his heart rate had become rapid and his heart beat unduly forceful.

He had a long history of episodes of fever, abdominal pain and vomiting which had been diagnosed as “abdominal migraine”. Both parents and his 4-year-old brother were healthy. His father was a factory worker and the family lived in a modern two-bedroomed flat.

On examination his weight was 30kg and his height was 138cm (growth charts Q11). He was alert and afebrile. His respiratory rate was 40/minute and his pulse rate was 130/minute. There was some pitting oedema over the dorsum of each foot. Jugular venous pressure was 5cm above the sternal angle. The apex beat was in the fifth interspace in the anterior axillary line and was thrusting in character. The first and second heart sounds were normal; the third heart sound was heard in the apical and left parasternal regions. The femoral pulses were readily palpable.

The blood pressure was 160/110 mmHg. Fine crepitations were heard at both lung bases. The appearance of the fundus is shown (Q10). The liver edge was palpable 3cm below the costal margin. Neither bladder nor kidneys could be palpated and there was no abdominal tenderness. Urinalysis was positive for protein (+) and negative for both glucose and blood.

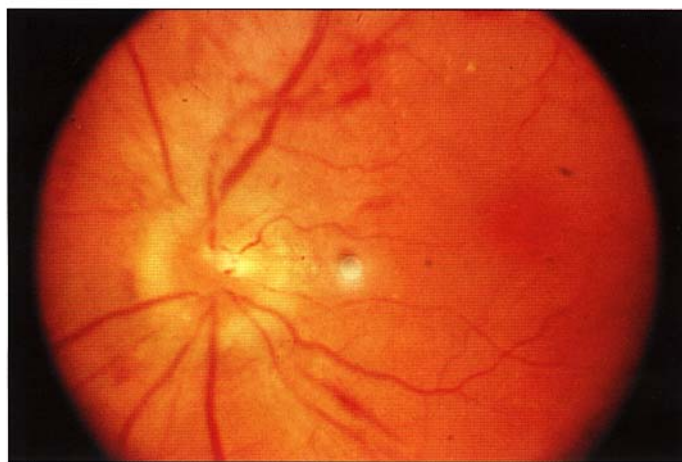
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Hb	9.2 g/dl
MCV	73 fl
MCH	23 pg
MCHC	31 g/l
WBC	$8.0 \times 10^9/l$
neutrophils	$5.20 \times 10^9/l$
lymphocytes	$2.64 \times 10^9/l$
monocytes	$0.08 \times 10^9/l$
eosinophils	$0.08 \times 10^9/l$
plasma sodium	133 mmol/l
plasma potassium	4.0 mmol/l
plasma chloride	97 mmol/l
plasma bicarbonate	20 mmol/l
plasma urea	18 mmol/l
plasma creatinine	300 μ mol/l
plasma total protein	70 g/l
plasma albumin	38 g/l
plasma calcium	2.1 mmol/l
plasma phosphate	2.7 mmol/l (normal range 0.99-1.57)
plasma alkaline phosphate	496 IU/l (normal range for age 71-234)
Chest x-ray	normal
Abdominal ultrasound:	Kidneys small with increased echogenicity No bladder abnormality

This question continues on the next page.

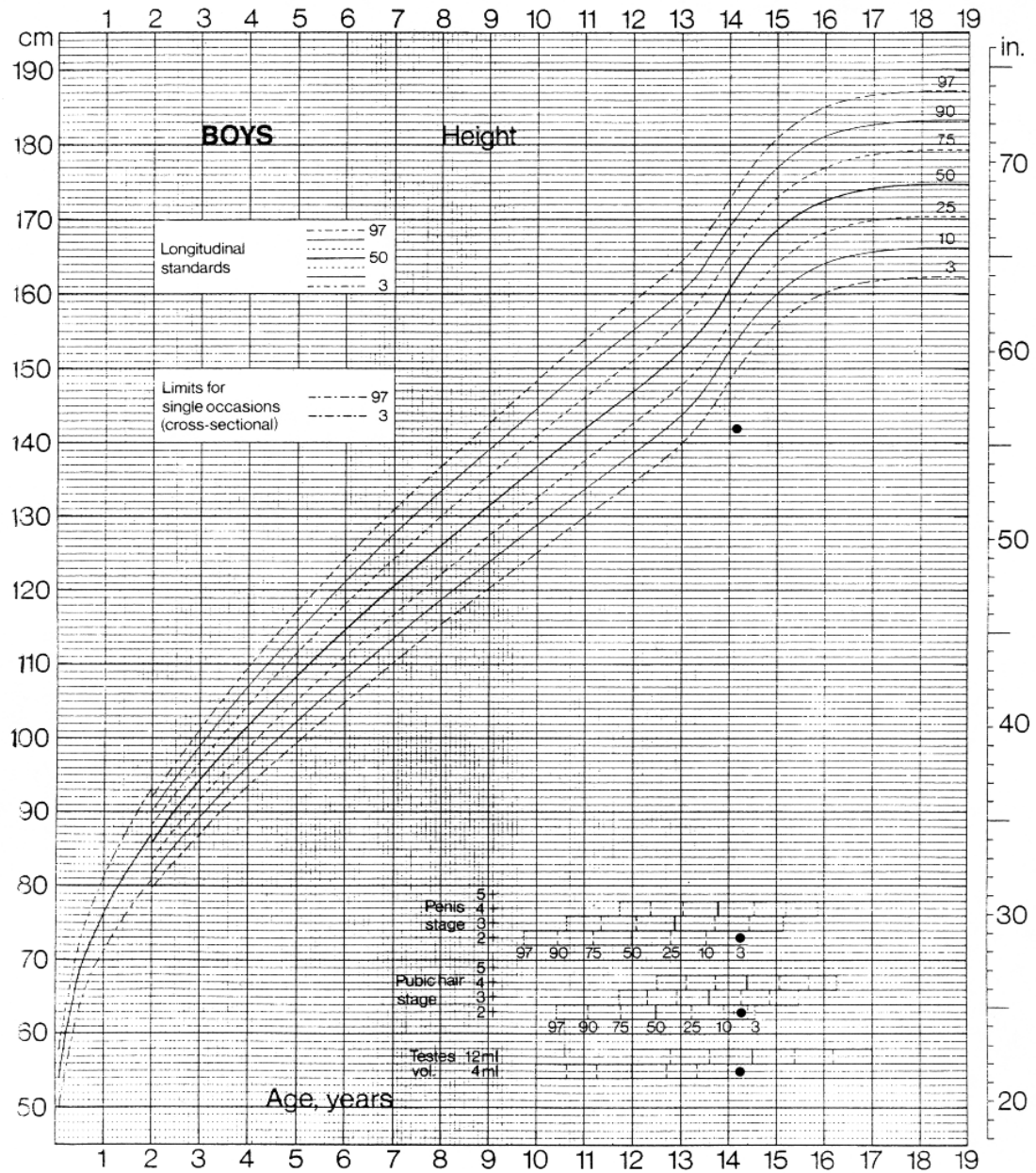


Q9



Q10

Q11



9. What is the most important abnormality on the radiograph of the hand shown in Q9? (1 mark)

SELECT ONE ANSWER ONLY

- A delayed bone age
- B osteomalacia
- C osteoporosis
- D splayed epiphyses
- E subperiosteal erosions

10. What is the most likely pathogenesis of the abnormality shown in Q9? (2 marks)

SELECT ONE ANSWER ONLY

- A chronic ill health
- B hypophosphataemia
- C poor dietary calcium intake
- D primary hyperparathyroidism
- E secondary hyperparathyroidism
- F vitamin D deficiency

This question continues on the next page.

11. What are the two most important features demonstrated on the growth chart? (3 marks)
(Q11)

SELECT TWO ANSWERS ONLY

- A bone age: advanced
- B bone age: delayed
- C bone age: normal
- D height: high
- E height: low
- F height: normal
- G pubertal staging: advanced
- H pubertal staging: delayed
- I pubertal staging: normal
- J weight for height: high
- K weight for height: low
- L weight for height: normal

This question continues on the next page.

- 12.** What is the best interpretation of the appearance of the optic fundus (Q12)? (3 marks)
(Ignore the central white spot which is an artefact)

SELECT ONE ANSWER ONLY

- A diabetic retinopathy
- B grade IV hypertensive retinopathy
- C optic atrophy
- D optic nerve head Drusen
- E pseudopapilloedema
- F traumatic retinal haemorrhage

- 13.** What is the most likely cause of his breathlessness? (2 marks)

SELECT ONE ANSWER ONLY

- A anaemia
- B left ventricular failure
- C metabolic acidosis
- D myocardial ischaemia
- E raised intracranial pressure
- F right ventricular failure

This question continues on the next page.

14. What is the most likely cause of his renal impairment? (2 marks)

SELECT ONE ANSWER ONLY

- A acute tubular necrosis
- B chronic glomerulonephritis
- C hypertensive nephropathy
- D hypovolaemia
- E reflux nephropathy

15. Which of the following renal investigations should now be performed? (2 marks)

SELECT TWO ANSWERS ONLY

- A abdominal CT
- B DMSA isotope scan
- C MAG 3 isotope scan
- D micturating cysto-urethrogram (MCUG)
- E renal arteriogram
- F renal biopsy

Questions 16, 17 and 18

- A acute lymphoblastic leukemia
- B beta thalassaemia major
- C beta thalassaemia trait
- D coeliac disease
- E congenital spherocytosis
- F Crohn's disease
- G haemolytic uraemic syndrome
- H immune haemolytic anaemia
- I Meckel's diverticulum
- J tuberculosis

Choose the one most likely diagnosis from the list above which best matches each of the following cases. **Note:** Each option may be used once, more than once or not at all.

SELECT ONE ANSWER ONLY FOR EACH QUESTION

- 16.** A previously well 2-year-old presents with a 3 week history of pallor, jaundice, lethargy and on examination has a spleen of 2 cms below the costal margin. The GP's full blood count shows Hb: 2.9gms white cell count $1.2 \times 10^9/l$, platelets 76, red cells indices normal. Lymphocytes 0.7, neutrophils 0.4.
- 17.** A normally grown 4-year-old Caucasian boy presents with lethargy and breathlessness of several weeks duration. On examination apart from pallor there is nothing else remarkable. Initial investigations have shown Hb 4 gm/dl. White cell count $7.0 \times 10^9/l$, platelets $520 \times 10^9/l$, reticulocytes 0.3%, blood film hypochromic/microcytic. Occasional atypical lymphocyte. ESR 25, ferritin 3.
- 18.** A 4-year-old presents with a 2-week history of lethargy and pallor following a viral illness. He looks slight jaundiced. Investigations show a Hb of 6.4 gms/dl, white cell count $12.5 \times 10^9/l$ (Neutrophils 5, lymphocytes 7, red cells 0.5), platelets $210 \times 10^9/l$. Blood film – MCV 90, occasional spherocytes, some atypical lymphocytes, no blast cells, reticulocytes count 6 %. Mycoplasma, IgM Positive.

Question 19

This is the face of a child aged three months with a normal platelet count who was otherwise well.

19. What is the best description of this lesion (3 marks)

SELECT ONE ANSWER ONLY

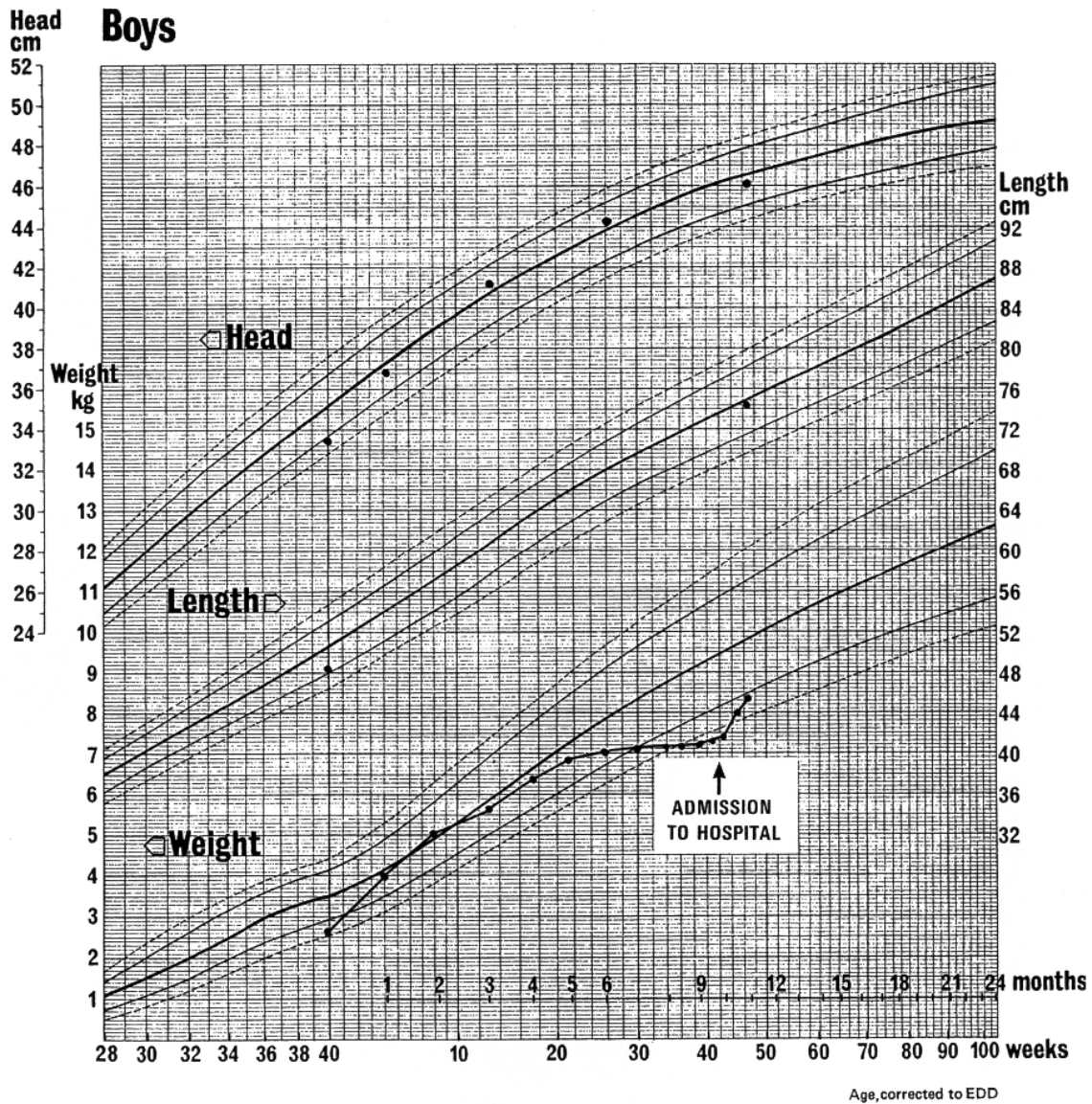
- A cavernous haemangioma
- B haemangioma
- C Kasabach-Merrit syndrome
- D mixed capillary/cavernous haemangioma
- E port wine naevus
- F cystic hygroma
- G strawberry naevus



Q19

Questions 20 and 21

This is the growth chart of a 10-month-old boy who was admitted to hospital because of vomiting and poor weight gain. The baby fed well on a mixed diet. The stools were normal. There were no abnormal findings apart from wasting.



Q20 & 21

This question continues on the next page.

20. What is the most likely diagnosis? (2 marks)

SELECT ONE ANSWER ONLY

- A coeliac disease
- B cow's milk protein intolerance
- C cystic fibrosis
- D gastro-oesophageal reflux disease
- E neglect

21. In the light of this growth chart, select the single most helpful assessment. (2 marks)

SELECT ONE ANSWER ONLY

- A case conference
- B measure antigliadin and anti-endomysial antibodies
- C oesophageal pH monitoring
- D RAST (radioallergosorbent test) for cow's milk IgE
- E sweat test

Questions 22 and 23

The following haematological picture was found in a girl aged 16 years in early pregnancy. She was born in the UK to parents of Afro Caribbean origin, who have been very supportive of her during this unexpected pregnancy. She has no previous medical history of note.

Hb	10.5 g/dl
RBC	$5.50 \times 10^{12}/l$
PCV	0.32
MCV	58 fl
MCH	20 pg
WBC	$9.2 \times 10^9/l$ (normal differential)
Platelet count	$250 \times 10^9/l$

Red cell appearances microcytic and hypochromic; many target cells

22. Select one further investigation which would elucidate the cause of the anaemia (2 marks)

SELECT ONE ANSWER ONLY

- A ferritin
- B haemoglobin electrophoresis
- C red cell folate level
- D bone marrow aspiration
- E serum B12 level

This question continues on the next page.

23. What is the most likely diagnosis? (3 marks)

SELECT ONE ANSWER ONLY

- A anaemia due to folate deficiency
- B beta thalassaemia
- C beta thalassaemia trait
- D chronic blood loss
- E iron deficient anaemia
- F normal variation

Question 24

The illustration shows the tongue of an otherwise healthy child. Her mother was worried about this appearance.

24. Which of the following is the most appropriate management? (3 marks)

SELECT ONE ANSWER ONLY

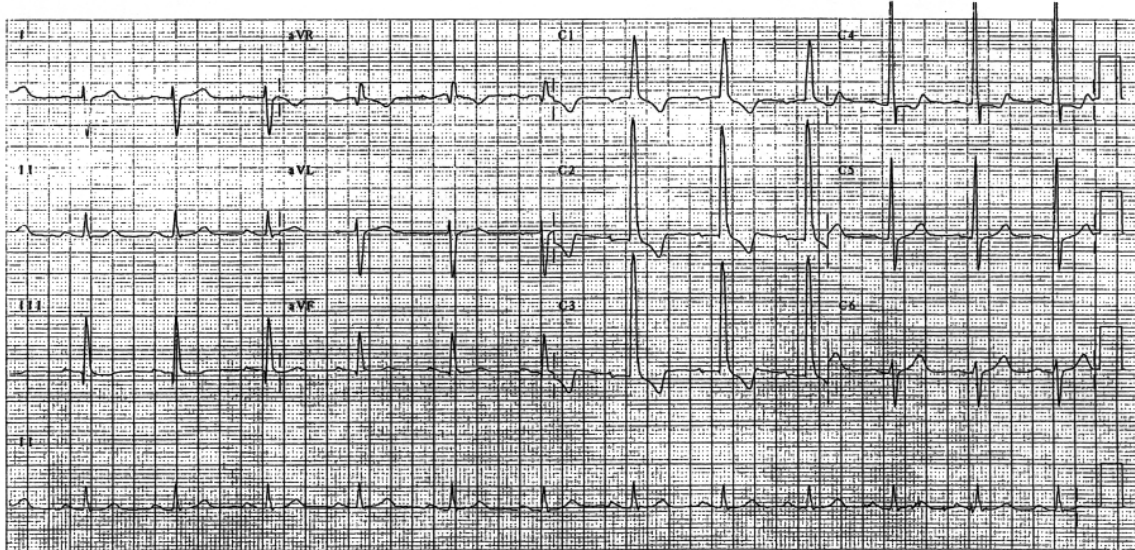
- A biopsy of the white area
- B determine HIV status
- C follow up for observation
- D nystatin
- E offer reassurance
- F serum iron/ferritin



Q24

Question 25

This is the ECG of a 15-year-old who had surgery for congenital heart disease in childhood. He was asymptomatic apart from breathlessness on vigorous exertion.



Q25

25. What two abnormalities are shown on this ECG? (4 marks)

SELECT TWO ANSWERS ONLY

- A complete left bundle branch block
- B complete right bundle branch block
- C dextrocardia
- D first degree heart block
- E inferoposterior myocardial infarction
- F left axis deviation
- G left ventricular hypertrophy and strain
- H partial left bundle branch block
- I right ventricular hypertrophy and strain
- J Wolff-Parkinson-White syndrome

Question 26

A 10-month-old boy presented with a history of failure to gain weight, constipation, polydipsia and polyuria.

Investigations:

blood glucose	4.8 mmol/l
plasma urea	2.7 mmol/l
plasma creatinine	54 µmol/l
plasma sodium	129 mmol/l
plasma potassium	2.6 mmol/l
plasma chloride	101 mmol/l
plasma bicarbonate	18 mmol/l
plasma calcium	2.59 mmol/l
plasma phosphate	0.85 mmol/l (1.1-1.9 mmol/l)
plasma alkaline phosphatase	270 IU/l (normal range 71-212 IU/l)
plasma amino acids	All quantitatively normal
pH	7.36 (hydrogen ion concentration 44 nmol/l)
Base deficit	-8.2 mmol/l

Urine analysis (24 hour collection)

volume	753mls on fluid intake 965mls
pH	7.0
glucose	12 mmol/l
sodium	25 mmol/l
potassium	16 mmol/l
urea	60 mmol/l
creatinine	0.4 µmol/l
phosphate	6.64 mmol/l
calcium	0.5 mmol/l
gross generalised amino-aciduria	
Ultrasound of kidneys and urinary tract	normal
X-ray of skeleton	slight demineralisation but no other abnormality

This question continues on the next page.

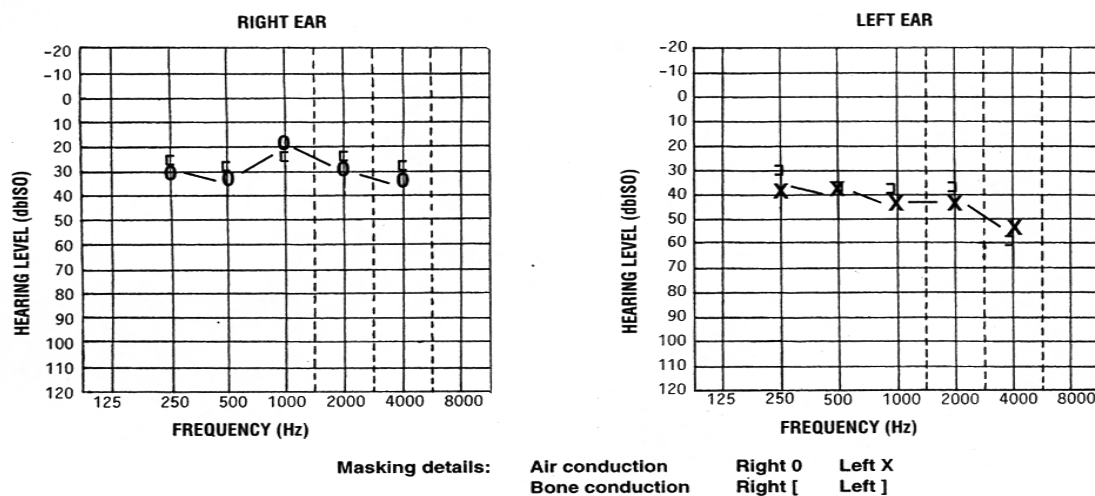
26. Choose three disturbances of renal physiology other than the amino-aciduria which have been demonstrated by these data. (4 marks)

SELECT THREE ANSWERS ONLY

- A acute renal failure
- B chronic renal failure
- C diabetes mellitus
- D excess hydrogen ion excretion
- E excess phosphate excretion
- F excess sodium excretion
- G inadequate phosphate excretion
- H inadequate potassium excretion
- I inadequate sodium excretion
- J renal glycosuria

Question 27

This is the audiogram of a 7-year-old child.



Q27

27. What abnormality is revealed by this audiogram? (5 marks)

SELECT ONE ANSWER ONLY

- A bilateral moderate conductive hearing loss
- B bilateral moderate sensorineural hearing loss
- C bilateral severe conductive hearing loss
- D bilateral severe sensorineural hearing loss
- E left moderate conductive hearing loss
- F left moderate sensorineural hearing loss
- G left severe conductive hearing loss
- H left severe sensorineural hearing loss
- I right moderate conductive hearing loss
- J right moderate sensorineural hearing loss
- K right severe conductive hearing loss
- L right severe sensorineural hearing loss

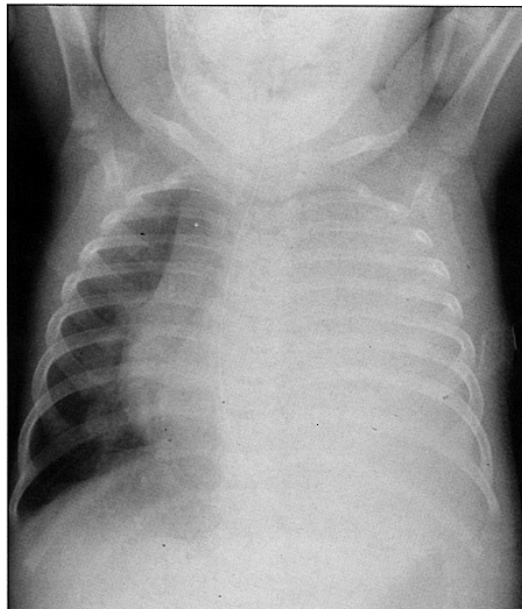
Question 28

This 2-year-old boy presented with a two weeks history of cough, fever and increasing breathlessness.

28. What are the two most important abnormalities present? (4 marks)

SELECT TWO ANSWERS ONLY

- A left lower lobe consolidation
- B left pleural effusion
- C left upper and lower lobe consolidation
- D left upper lobe consolidation
- E mediastinal shift
- F right lower lobe consolidation
- G right middle lobe consolidation
- H right pleural effusion
- I right upper lobe consolidation
- J right widespread consolidation



Q28

Questions 29, 30 and 31

- | | | | |
|---|----------------|---|------------------|
| A | ACTH | I | Lorazepam |
| B | Carbamazepine | J | Paraldehyde |
| C | Clonazepam | K | Phenobarbitone |
| D | Diazepam | L | Phenytoin |
| E | Ethosuximide | M | Sodium valproate |
| F | Gabapentin | N | Topiramate |
| G | Ketogenic diet | O | Vigabatrin |
| H | lamotrigine | P | None |

The above drugs are used for the treatment of epilepsy.

Which drug would you use as a first line treatment in the following cases?

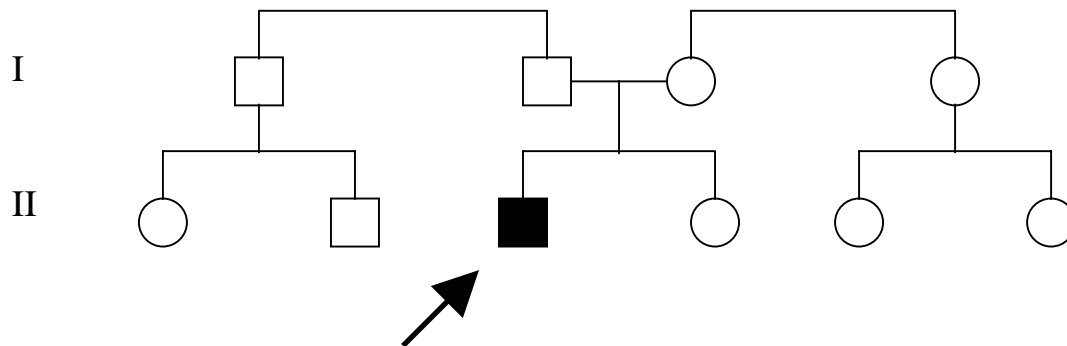
Note: *Each option may be used once, more than once or not at all.*

SELECT ONE ANSWER ONLY FOR EACH QUESTION

- 29.** A term newborn with birth asphyxia presents at 12 hours of age with frequent short-lived generalised convulsions.
- 30.** A 16-year-old girl doing her GCSE exams with generalised tonic epilepsy.
- 31.** A 7-year-old boy with generalised absence seizures.

Questions 32 and 33

This index child possesses the karyotype 47, XXY. His parents, aged 25 and 26, have normal karyotypes.



32. What is the degree of risk of his parents having subsequent children with this condition? (2 marks)

SELECT ONE ANSWER ONLY

- A zero
- B less than 1%
- C 1%
- D 5%
- E 25%
- F 50%
- G 67%
- H 100%

This question continues on the next page.

33. What is the likelihood of the index child having affected offspring? (2 marks)

SELECT ONE ANSWER ONLY

- A zero
- B less than 1%
- C 1%
- D 5%
- E 25%
- F 50%
- G 67%
- H 100%

Question 34

34. This is the face of a boy aged five years.

What is the most likely diagnosis? (3marks)

SELECT ONE ANSWER ONLY

- A eczema herpeticum
- B herpes simplex
- C impetigo
- D Stevens Johnson syndrome
- E toxic epidermal necrolysis



Q34

END OF SPECIMEN PAPER 1

Paper One Web Specimen Paper - Answer Key			
Question Number	Answer Key	Answer and Description	Marks
1	B	nasopharyngeal aspirate for respiratory viruses	3
2	F	viral bronchiolitis	4
3	A	left sided Horner's syndrome	3
4	B	partially compensated respiratory acidosis	2
5	B	ensure nasal CPAP circuit is working correctly	3
6	C	CT scan	3
7	E	administer intravenous mannitol	2
	G	arrange for intubation and ventilation	2
8	E	subarachnoid haemorrhage	3
9	E	subperiosteal erosions	1
10	E	secondary hyperparathyroidism	2
11	E	Height: low	1
	H	pubertal staging: delayed	2
12	B	grade IV hypertensive retinopathy	3
13	B	left ventricular failure	2
14	E	reflux nephropathy	2
15	B	DMSA isotope scan	1
	D	micturating cysto-urethrogram (MCUG)	1
16	A	acute lymphoblastic leukaemia	3
17	F	Crohn's disease	3
18	H	immune haemolytic anaemia	3
19	A	cavernous haemangioma	3
20	E	neglect	2
21	A	case conference	2
22	B	haemoglobin electrophoresis	3
23	C	beta thalassaemia trait	2
24	E	offer reassurance	3
25	D	first degree heart block	2
	I	right ventricular hypertrophy and strain	2
26	E	excess phosphate excretion	1
	F	excess sodium excretion	1
	J	renal glycosuria	1
27	B	bilateral moderate sensorineural hearing loss	5
28	B	left pleural effusion	2
	E	mediastinal shift	2
29	K	Phenobarbitone	3
30	H	Lamotrigine	3
31	M	Sodium valproate	3
32	B	less than 1%	2
33	A	zero	2
34	D	Stevens Johnson syndrome	3